A COMPREHENSIVE STUDY ON CRANIOVERTEBRAL JUNCTION ANOMALIES

Dissertation submitted in partial fulfillment of the requirements for the degree

Of

M.Ch., Branch – II (3 Years)

NEUROSURGERY

THE TAMILNADU DR. M.G.R. MEDICAL UNIVERSITY

INSTITUTE OF NEUROLOGY

MADRAS MEDICAL COLLEGE, CHENNAI – 600 003.

AUGUST 2014

CERTIFICATE

This is to certify that this dissertation entitled **"A Comprehensive study on Craniovertebral Junction Anomalies"** submitted by **Dr.D.R. Shankar**, appearing for **M.Ch (Neurosurgery)** degree examination in August 2014 is a original bonafide record of work done from January 2012 to December 2013 by him under my guidance and supervision in partial fulfillment of requirement of the Tamil Nadu Dr.M.G.R. Medical University, Chennai. I forward this to the Tamil Nadu Dr.M.G.R. Medical University, Chennai, Tamil Nadu, India.

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DECLARATION

I solemnly declare that this dissertation **"A COMPREHENSIVE STUDY ON CRANIOVERTEBRAL JUNCTION ANOMALIES"** was prepared by me in the Institute of Neurology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai-3, under the guidance and supervision of Professor of Neurosurgery, Institute of Neurology, Madras Medical College and Rajiv Gandhi Government General Hospital, Chennai from January 2012 to December 2013.

This dissertation is submitted to the Tamilnadu Dr. M.G.R Medical University, Chennai in partial fulfillment of the University requirements for the award of degree of M.Ch., Neurosurgery.

Place : Chennai

Date : 28.03.2014

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I specially thank my fellow colleagues who helped me in this study. I thank all my patients and their relatives who have participated in this study.

INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI-3

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CERTIFICATE OF APPROVAL

To
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Dear Dr. D.R. Shankar,

The Institutional Ethics Committee of Madras Medical College, reviewed and discussed your application for approval of the proposal entitled **"A Comprehensive study on Craniovertebral Junction Anomalies"** No.38032014

The following members of Ethics Committee were present in the meeting held on 11.03.2014 conducted at Madras Medical College, Chennai-3.

1.	Dr. C. Rajendran, M.D.	-- Chairperson
2.	Dr. R. Vimala, M.D. Dean, MMC, Ch-3.	-- Deputy Chairperson
3.	Prof. Kalaiselvi, MD Vice-Principal, MMC, Ch-3	-- Member Secretary
4.	Prof. Nandhini, M.D. Inst. of Pharmacology, MMC, Ch-3.	-- Member
5.	Prof. Bhavani Shankar, M.S. Prof & HOD of General Surgery, MMC, Ch-3.	-- Member
6.	Prof. V. Padmavathi, M.D. I/c Director of Pathology, MMC, Ch-3.	-- Member
7.	Thiru. S. Govindasamy, BABL	-- Lawyer
8.	Tmt. Arnold Saulina, MA MSW	-- Social Scientist
9.	Thiru. S. Ramesh Kumar, Administrative Officer, MMC, Ch-3.	-- Layperson

We approve the proposal to be conducted in its presented form.

Sd/Chairman & Other Members

The Institutional Ethics Committee expects to be informed about the progress of the study, and SAE occurring in the course of the study, any changes in the protocol and patients information / informed consent and asks to be provided a copy of the final report.

13/3/14

Member Secretary, Ethics Committee

MEMBER SECRETARY
INSTITUTIONAL ETHICS COMMITTEE
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CONTENTS

ABSTRACT

OBJECTIVES ;

To Analyse comprehensively about the incidence of Craniovertebral Junction Anomalies in different age groups, their sex prevalence, their clinical findings, radiological abnormalities, the different surgical procedures done and their final outcome.

MATERIALS AND METHODS :

About 50 patients with CVJ Anomalies were taken for this prospective study. All the Congenital& Acquired CVJ Anomalies in all the sex groups were taken for the study. The incidental Arnold Chiari malformation was excluded from this study. All the personal, clinical, radiological & surgical treatment and the final outcome were entered and analysed.

RESULTS :

The CVJ Anomalies are more common in young adults (28%), almost equal in both sexes. The Congenital (68%) are more common than the Acquired. Atlantoaxial Dislocation (22%) is the most common congenital bony anomaly and Arnold Chiari malformation is the most common soft tissue anomaly. The patients with increased ADI 3 to 5mm showed 77% improvement after surgery.

CONCLUSION ;

The Atlantodental Interval is the important pre operative prognostic marker. The Craniometric lines arew very arbitrary. No definite surgical technique is correlated with the post operative surgical outcome.

INTRODUCTION

Craniovertebral Junction, being the transit zone between cranium and spine, is the most complex and dynamic region of the cervical spine. It has complex bony anatomy and intricate tissues and major neurovascular structures. The subject of CVJ is under discussion and evaluation over a century and numbers of classical reviews have attempted to clarify a variety of complex associated issues.

The incidence of different types of CCVJ Anomalies varies with demographic environment &ill-defined genetic factors. CVJ Anomalies are more frequently found in Indian subcontinent than anywhere else in the world. Even in India, these anomalies are more frequently documented from Bihar, Uttar Pradesh, and Rajasthan& Gujarat. The reason for this geographical clustering is more speculative. The CVJ Anomalies can be either due to Bony or Soft tissueAnomalies. They are common in all age groups and almost equal in both sex groups.The anomalies can be due to congenital and Acquired causes. There has been a renewed interest in the normal anatomy & pathological lesions of CVJ Anomalies with Dynamic X rays, CT &MRI.The clinical features are often delayed upto 2nd or 3 rd decade, since they are subtle and often missed.The surgical management of CVJ Anomalies is complex due to the relative difficulty in accessing the region,critical Neurovascular structures and the intricate Biomechanical issues involoved. Inspite of various

surgical procedures, the commonly done procedures are Foramen Magnum Decompression, C1 C2 wiring, C1 lateral mass & C2 pedicle screw fixation and Trans Oral Odointoidectomy. The surgical indications for each approach are still under discussion.

The management protocol varies with each patient and there are several studies conducted on this issue, trying to arrive at a consensus.

Hence an attempt was made to analyse comprehensively on the incidence of CVJ anomalies among various age groups, its various clinical features, different radiological investigations, various etiological diagnosis, different surgical procedures and it's final surgical outcome.

AIMS AND OBJECTIVES

The Aims & Objectives of this study are as follows;

1. To study the incidence of various Bony & Soft tissue CVJ Anomalies.
2. To study the different clinical features of various CVJ Anomalies.
3. To analyse the various etiological factors contributing to CVJ Anomalies.
4. To study the different radiological Investigations & their correlation with the final surgical outcome.
5. To analyse the various surgical procedures done for CVJ Anomalies&its final surgical outcome.

REVIEW OF LITERATURE

Literature is reviewed as follows ;

1. Outline of Craniovertebral Junction anatomy.
2. Etiological Classification of different CVJ Anomalies.
3. Various Clinical presentations of CVJ Anomalies.
4. Different Radiological Investigations to diagnose the CVJ Anomalies.
5. Various Surgical Treatment Options.
6. Brief view of Literature on CVJ Anomalies.

1. OUTLINE OF ANATOMY OF CRANIOVERTEBRAL JUNCTION

The Atlantoaxial complex is unique among the intervertebral joints. And it is horizontally oriented. The facet joints are relatively flat. It also allows a pivoting motion at the atlantodental articulation, which is supported by the special ligamentous support.

The second cervical nerve exits from the cervical canal immediately adjacent and dorsal to the joint capsules. The transverse atlantal ligament is a band 3 to 5 mm thick that originates from the tubercles and the inner aspect of

the lateral masses of the atlas vertebra and is in close apposition to the Odontoid.

By itself, the geometry of the Craniovertebral complex is meant to provide mobility at the cost of stability.

Stability of this joint complex is contributed by the ligaments. Also it is by the little contribution from bony articulations and joint capsules.

1. **Ligaments that connect the atlas to the Occiput :**

A. Anterior Atlanto Occipital membrane:

It extends from anterior margin of foramen Magnum to anterior arch of C1. It is the cephalad extension of Anterior Longitudinal Ligament

B. Posterior Atlanto Occipital membrane :

It connects the posterior margin of Foramen Magnum to the posterior arch of C1.

C. It is the Ascending band of Cruciate ligament.

2. **Ligaments that connect Axis to the Occiput :**

A) Tectorial Membrane:

Superficialpart: It is the Cephalad continuation of Posterior longitudinal ligament.A strong band connecting the dorsal surfaceof

Foramen Magnum above and the dorsal surface of C2 & C3 bodies below.

Deep portion : It is located laterally, connects Axis to the occipital condyles.

1. Ligaments that connect Axis to Atlas ;
2. A)Transverse Ligament : It is the horizontal component of Cruciate ligament.
3. It provides the majority of strength.
4. B)Atlanto-alar portion of Alar ligament
5. C)Descending band of Cruciate ligament.

II. CLASSIFICATION OF CVJ ANOMALIES ;

I. Congenital Anomalies ;

A. Malformations of Occipital Bone :

a. Clivus segmentations
b. Remnants around the Foramen magnum
c. Atlas variants
d. Dens segmentation anomalies.

B. Malformations of Atlas :

a. Assimilation of Atlas
b. Atlantoaxial fusion
c. Aplasia of Atlas arches.

C. Malformations of the Axis :

a. Irregular AtlantoAxial segmentation

b. Dens Dysplasias

a. Ossiculum terminale persistens

b. OsOdontoideum

c. Hypoplasia – aplasia

d. Segmentation failure of C2\C3

II. Developmental and Acquired abnormalities craniocervical Junction

A. Abnormalities of Foramen Magnum

1. Secondary Basilar Invagination (Basilar Impression) : Paget's disease, Rheumatoid Arthritis, Osteomalacia, Rickets.
2. Foraminal stenosis (e.g., Achondroplasia)

B. Atlantoaxial instability

1. Errors in metabolism (e.g., Morquio's syndrome)
2. Down syndrome
3. Infections (e.g., Grisel's syndrome)
4. Inflammatory (e.g., Rheumatoid arthritis)
5. Traumatic Occipitoatlantal and atlantoaxial dislocation, Osodointoideum
6. Tumors (e.g., Neurofibromatosis, syringomyelia)
7. Miscellaneous (e.g., fetal warfarin syndrome, Conradi's syndrome)

THE ARNOLD CHIARI MALFORMATIONS

CHIARI TYPE	FEATURES
1	Tonsillar Herniation >5mm inferior to foramen magnum plane. No associated brainstem herniation
2	Herniation of cerebellar vermis, brainstem and fourth ventricle through foramen magnum. Associated myelomeningocele and multiple brain anomalies. Hydrocephalus &syringomyelia very common.
3	High cervical or occipital encephalocele
4	Hypoplasia or aplasia of cerebellum and tentorium cerebelli.

II. CLINCAL FEATURES ;

The most interesting feature is the diversity of clinical feature as a result of compromise of lower brainstem, cervical spinal cord, cranial nerves, cervical roots and vascular supply.

The congenital Anomalies are associated with abnormal physical appearancelike Head tilt, short neck, low hair line, limited neck movements (Klippel-Feil syndrome)

There is an increased incidence of association with Achondroplasia, Spondyloephiphyseal dysplasias and dwarfism.

The most frequent symptom is the suboccipital neck pain, radiating to the cranium. The features of myelopathy may present in the form of monoparesis, hemiparesis, paraparesis & ascending pattern of Quadriparesis. Central cord syndrome is seen in children with Basilar Invagination.

Sensory abnormalities are manifested as neurological deficits related to the anterolateral and posterior columns dysfunction.

Brainstem and cranial nerve deficits can cause abnormalities like dysphagia, nasal regurgitation, change in voice, loss of facial sensation, sleep apnoea.

Downbeat Nystagmus is a cardinal sign, more characteristically seen with strictly compressive CVJ lesions.The excessive mobility of an unstable CVJ can cause trauma to the Anterior spinal artery and the perforating vessels of the upper cervical cord and medulla oblongata producing features of Vertebrobasilar Insufficiency like vertigo, syncopal attacks, transient loss of consciousness.

RADIOLOGICAL INVESTIGATIONS :

1) Dynamic Xrays Cervical spine:

The X rays cervical spine, Lateral views both Flexion and Extension views to assess the reducibility of the CVJ Anomaly. Atlanto Dental Interval (ADI) – the distance from the anterior surface of the Dens to the posterior edge of the anterior arch of Atlas.

Normal values : Adults – 3mm.Children – 4mm.

2) CT Cranio vertebral Junction:

To assess the bony anomalies like, Assimilation of Atlas, Defects in the arches of Atlas, Atlanto Axial Subluxation, Basilar Invagination, Platybasia, & traumatic Fractures.

To look for associated skeletal deformities like kyphosis, scoliosis. Congenital Block vertebrae are well made out.

To study the rotatory atlantoaxial subluxation, focal hematomas, joint incongruity.

To look for bony erosion, displacement in inflammatory conditions like Tuberculosis, Rheumatoid arthritis.

3) MRI Cervical Spine:

To study the tonsillar descent, Brainstem herniation, Spinal canal diameter, cord compression, presence of syrinx and intrinsic cord changes.

CraniometricAssessment:

Various anatomical lines are studied at the level of Foramen Magnum in X rays, CT scan & MRI Cervical spine. Not a single line is used.

Mc Rae's line – it is the foramen magnum line connecting the Basion to Opisthion. The normal foramen magnum diameter is 40 mm. The effective canal diameter less than 20 mm implies foraminal stenosis with severe cervico medullary compression.

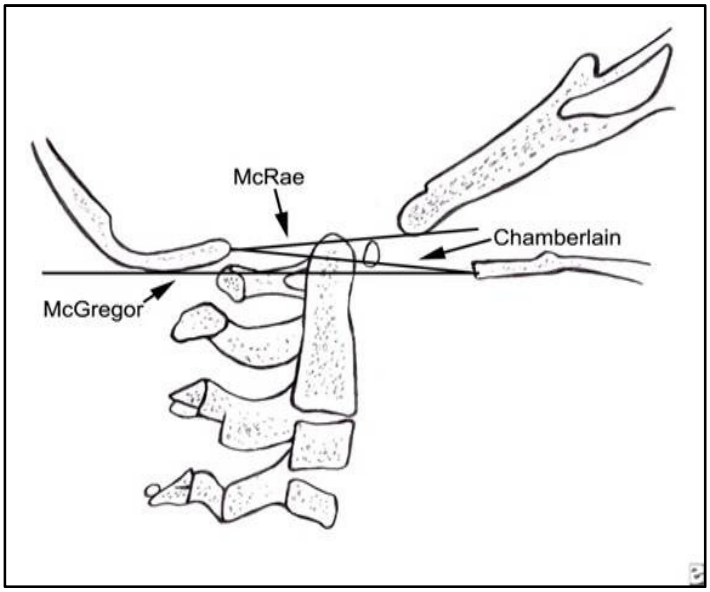
Mc Gregor's line (Palato suboccipital line) – it is the line connecting the posterior most surface of hard palate to the internal surface of the Occiput. The tip of the Odontoid process should lie either below this line.

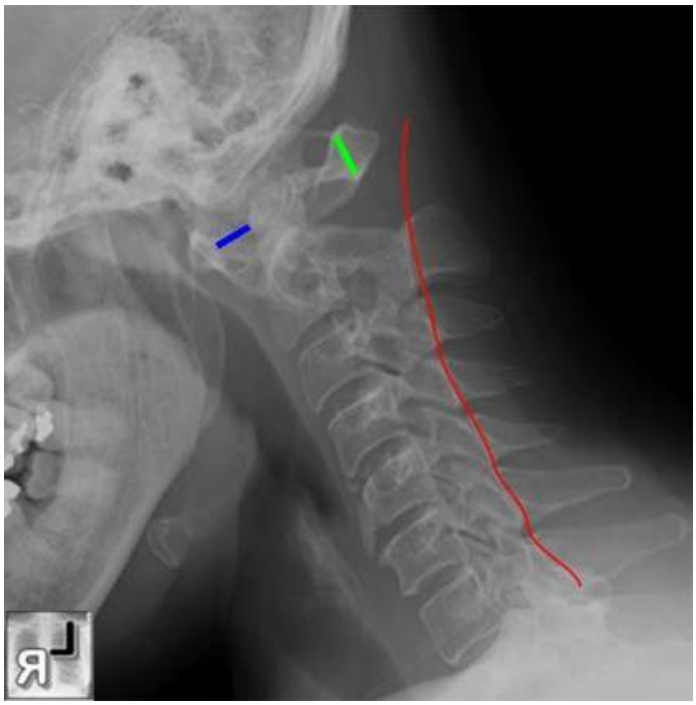
Chamberlain's line (Palato occipital line) – it is the line connecting the posterior surface of hard palate to the inferiormost surface of theOpisthion. Normally the tip of odontoid should lie below this line.

Wackenheim's Clivus canal line - it is the tangential line along the clivus & by extrapolating it inferiorly downwards. Normally the Odontoid process lies below or does not cross more than 2.5mm above this line.

Welcher's Basal angle – it is the angle formed between the Nasion tuberculum line and the tuberculum basion line. Normally it is around 130 degrees. More than 130 degrees implies flat skull base.

CRANIOMETRIC LINES AT CV JUNCTION





Case no. 13 with increased Atlantodental Interval (>3mm)

TREATMENT OPTIONS

All the reducible CV Junction Anomalies are better treated with surgical modality. The irreducible anomalies are immobilized with Philadelphia collar or halo brace and managed conservatively.

The various surgical treatment options :

1. Foramen Magnum Decompression :
2. It is done by the Suboccipital craniectomy and removal of posterior arch of Atlas. Most commonly done in Arnold Chari Malformations.
3. C1 C2 Wiring: C1 & C2 are fixed by Sublaminar titanium wires by Gallies fusion, Brooke's fusion and Sonntag fusion techniques.
4. C1 C2 Fixation: Commonly done by C1Lateral mass & C2 pedicle screw fixation by Harms Technique, Transarticular screws by Maegerl's technique, and Translaminar screw fixation in suspected vertebral artery anomalous course with fear of vertebral artery injury.
5. OccipitocervicalFusion

   It is done by stainlesssteel or titanium Contoured rod or loop with screw fixation. This technique is gaining popularity nowadays , because of better post-operative outcome.
6. Transoral Odointoidectomy :

   Transoral excision of Odontoid is done by que, transpalatal, transpharyngeal routes, also by transnasal endoscopic route to relieve the ventral compression.

BRIEF REVIEW OF LITERATURE

Atul Goel et al had conducted a retrospective study in about 3300 patients from 1971 to 2009 and divided the Basilar Invagination into two groups based on presence (Group A) or absence (Group B) of clinical & Radiological evidence of instability.The pathogenesis and clinical features in Group A Basilar Invagination appeared to be related to mechanical instabilitywhere as Group B related to be secondary to embryological dysgenesis. Group A patients treated by Facetal distraction & Lateral mass fixation, whereas Foramen Magnum Decompression was done in Group B patients.

AbrarAbadtrani et al studied CVJ Injuries in about 83 patients from 2003 to 2008.The classical clinical features described are the pyramidal signs, movement restriction, Facial or Hand Asymmetry, Torticollis etc., Early surgical intervention (i.e. within 2 weeks) carries good prognosis.

Nicolia Di Lorenzo et al had conducted a retrospective study in 63 patients from 1953 to 1979 to assess the efficacy of treatment in relieving symptoms associated with CVJ Anomalies. About 58 patients underwent Posterior cervicomedullary Decompression & others underwent Transoral Clivus-Atlanto Odointoidectomy. Long term results with posterior approach

showed that 50% patients benefitted by surgery, 25% remain unchanged, 25% continued to be deteriorated.

Anterior Decompression is preferable to a posterior approach, only if the CVJ involves a ventral deformity in the absence of dorsal compression by soft tissues.

Gyo-chang song et al had studies retrospectively in 82 patients from 2005 to 2011about the clinical outcome & effectiveness of Occipitocervical fusion as the surgical treatment of CVJ Instability. About 9 patients remained the same after surgery. Among 73 patients with cervical myelopathy, clinical Improvement noted in 58 cases, 3 patients died about 2 months after surgery, Fusion achieved in about 47 patients.

Sanjay Behari et al had conducted retrospective study on 54 patients from 2000 to 2006 who underwent Occipito cervical Contoured rod stabilization. About 50 patients had AAD, 3 had CVJ Tuberculosis, one had Rhematoid Arthritis. About 24 patients had improved, 18 patients got stabilized and 6 deteriorated at a mean follow up. About 6 patients had lost follow up.

S.S. Kale & Parkanj et al had studied retrospectively the CVJ Anomalies in consecutive 189 patients from 2001 to 2010. About 162 patients had developmental anomalies, 18 patients had traumatic causes, post

inflammatory due to Tuberculosis in 9 patients. Surgical procedures include Transoral Decompression (118), Occipitocervical Fusion in 71 patients, contoured stainless rods &Sublaminar wires in 86 patients. Foramen Magnum Decompression done in 5 patients with Arnold Chiari Malformation, posterior decompression with posterior fixation done in 26 patients.

Giussani . C et al had studied a series of severely symptomatic CVJ Anomalies in children and analysed the chronological long term effectiveness of aggressive management of CVJ Anomalies in terms of clinical improvement, spinal stability & growth.

He had noticed the 3 Down Syndrome patients, 1 with Morquio syndrome & 1 with OsOdontoideum.

Atul Goel et al had studied the importance of Atlantoaxial joint distraction for Basilar Invagination patients with Rheumatoid Arthritis (59 patients) in whom the facets are osteoporotic and unsuitable for screw fixation. Manual distraction of facets of Atlas & axis and forced impaction of titanium spacer in the Joint in addition to the bone graft harvested from Iliac crest. Mean follow up period was from 2 to 24 months. All the patients showed symptomatic improvement & partial restoration of Craniovertebral alignment.

Arjun Shetty et al had studied retrospectively, the various Reduction Techniques in AtlantoAxial subluxation in about 63 years over 5 years,

including the Atlantoaxial Joint space release and a variety of manipulation procedures. About 49 had achieved optimum Reduction and 14 patients had near Normal Optimal reduction.

Wang C had studied the Intra operative reduction, Instrumentation & Fusion of CVJ Anomalies in about 33 patients. OsOdointoideum was seen in about 8 patients, Occipitalisation of Atlas in 19 patients, malunion of Odontoid in 5 patients.

Menezes et al had studied prospectively in 100 patients with primary CVJ Anomalies and hindbrain herniation syndrome. Hindbrain herniation seen with 4 the occipital sclerotome abnormalities, become symptomatic with canal diameter <19 mm. Ventral decompression relieved brainstem & cerebellar syndrome signs. Post operative Cine MRI shows reversal of Craniospinal CSF dissociation after ventral CVJ Decompression.

Stevens JM, Balper.C had conducted a prospective study on abnormalities of Odontoid process associated with Atlantoaxial Dislocation and Neurological instability in about 79 patients from 2001 to 2006 and concluded thatOs Odontoid is the prime congenital anomaly of Odontoid leading on to Atlantoaxial dislocation.

MATERIALS AND METHODS

About 50 patients with Craniovertebral Junction Anomalies who were treated in the Institute of Neurology, Rajiv Gandhi Government General Hospital, Chennai during the period of January 2012 to December 2013 have been studied prospectively.

Inclusion criteria for this study are as follows :

1. Bony CVJ Anomalies.
2. Soft tissue Anomalies.
3. All Age groups.

Exclusion criteria for this study are as follows :

1. Incidental Arnold Chiari Malformation.
2. Patients with Irreducible CV Junction Anomalies.
3. Patients who are not willing for surgery.
4. Patients who have lost follow up.

This is a prospective analytical study and all the patients in the study were entered into a proforma as given in Appendix I.

All the clinical, radiological,surgical treatment & outcome details were entered into the proforma included in the Appendix – 1.

CRITERIA FOR DETECTING THE INCIDENCE ;

All the patients personal details like name, age, sex were entered into the proforma.

CLINICAL CRITERIA FOR THE ANALYSIS OF SYMTOMATOLOGY ;

The patients symptoms of sensorimotor disturbances, cranial nerve disturbances, features of increased intracranial hypertension, autonomic disturbances, symptoms of vertebrobasilar insufficiency (vertigo, syncope) were analysed.

CLINICAL CRITERIA FOR THE DIAGNOSIS ;

After complete clinical examination, the sensory level, the motor level and the reflex level was found and the diagnosis of CVJ anomalies were attained. The other associated clinical features like Head tilt, short neck, webbed neck, Downbeat Nystagmus,Cranial nerve deficits, features of congenital syndromes like Down syndrome, Morquio syndrome, Klippel – Feil syndrome were taken into account.

RADOLOGICAL CRITERIA FOR THE DIAGNOSIS ;

X ray Cervical spine :

a. Occipitalisation of Atlas due to the assimilation of Atlas to the Occiput.

b. Defect in the arches of Atlas.

c. Atlanto Dental Interval (ADI) : It is the interval between posterior surface of Anterior Arch and the Anterior surface of Atlas. In adults, ADI >3mm & in children, ADI > 4mm was taken as positive for Atlanto Axial Subluxation.

d. Block Vertebrae – Congenital fusion of vertebral bodies.

Dynamic Xrays CVJ :

Both Flexion & Extension Lateral views of CVJ were viewed.

The increased ADI on Flexion view implies irreducible Atlanto Axial Subluxation.

Craniometric Assessment :

Various anatomical lines were studied from X Ray CVJ, CT scan CVJ and MRI CVJ.

1. **Mc Rae's line :** It is the foramen magnum line connecting the Basion and Opisthion.
2. **Mc Gregor line :** It is the line connecting the posterior surface of palate to the Occiput
3. **Chamberlain line :** It is the line connecting the posterior surface of palate to the inferior surface of the Occiput.

4. **Wackenheim's Clivus canal line :** It is the tangential line drawn along the clivus & by extrapolating it downwards. Normally the Odontoid should lie below or should not exceed 2.5mm above this line.
5. **Welcher's Basal Angle :** It is the Angle between the nasion tuberculum line & tuberculum Basion line. The angle above 130 degreewas taken as positive for Basilar Invagination.
6. **Powers ratio :** It is the ratio between the lines connecting Basion to anterior edge of posterior arch of Atlas and the line connecting the Opisthion to the posterior surface of Anterior arch of Atlas. Normal : 0.77. Abnormal values (>0.77) implies atlanto occipital Dislocation.

CT scan CV Junction :

Various Bony anomalies like defects in the arches of Atlas, Assimilation of Atlas, Block Vertebrae, Basilar Invagination, Atlanto Occipital Dislocation, Odontoid fracture were studied.

MRI CV Junction :

Apart from Craniometric assessment, Atlanto axial subluxation, tonsillar herniation, cerebellar descent, syringomyelia, intrinsic cord changes, prevertebral soft tissue collections were studied.

CRITERIA FOR ETIOLOGICAL DIAGNOSIS ;

Based on the history & thorough clinical examination, the CVJ Anomalies were broadly categorized into Congenital and Acquired.

Among the congenital Bony CVJ Anomalies, congenital Atlanto Axial subluxation, Basilar Invagination, Platybasia, Occipitalisation of Atlas, Defect in arches of Atlas were studied.

Among the Congenital Soft tissue CVJ Anomalies, Arnold Chiari malformations, Syringomyelia were studied. The associated features of congenital syndromes like Down syndrome, Morquio syndrome &KlippelFeil Syndrome were studied.

The Acquired CVJ Anomalies were studied under traumatic causes, Infective causes – Grisel Syndrome following Pharyngeal Infection, Inflammatory causes – following Tuberculosis, Rheumatoid arthritis, Neoplastic causes – following Foramen Magnum meningiomas, Neurofibromas.

CRITERIA FOR STUDYING THE SURGICAL TREATMENT ;

The patients who underwent the following surgeries were studied.

1. Formamen magnum Decompression.
2. C1 C2 Wiring using Stainless Steel or Titanium wires.
3. C1 Lateral mass & C2 pedicle screw fixation.

4. TransoralOdontoidectomy
5. Occipito Cervical Fusion.

CRITERIA FOR STUDYING THE OUTCOME ;

The patients were followed up at 1 month, 3 months, and 6 months. Thorough clinical examination was made and the outcome was measured based upon the improvement / deterioration / static neurological status by assessing the motor power by MRC Grade. Post operative Imaging was done to assess the Optimal reduction and restoration of Craniovertebral junction.

The AtlantodentalInterval (ADI 3-5mm, >5mm) was correlated with the final outcome of the patients and the statistical significance was found. The Craniometric lines were correlated with the final outcome and the statistical significance was found out.

The statistical significance was found in the patients, who underwent different surgical procedures by correlating with the outcome.

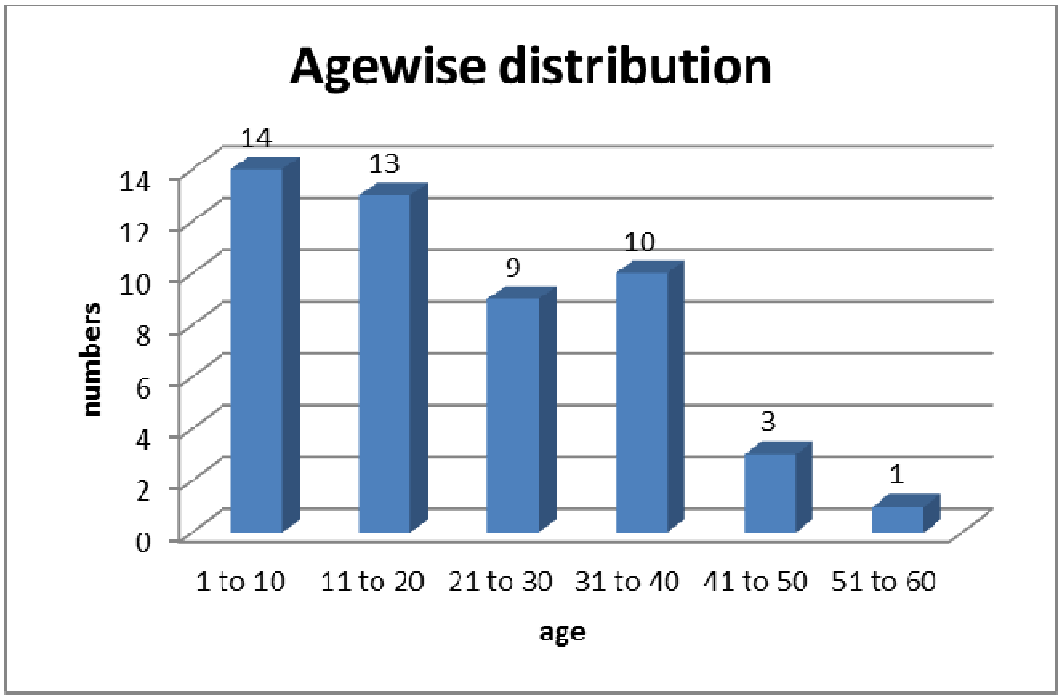
The Statistical Analysis was done by Chi-Square test by using Graph Pad Stat Software.

OBSERVATION AND RESULTS

About 50 cases of Craniovertebral junction Anomalies treated during the period of January 2012 to December 2013 were studied. The summary of the 50 cases is given in the Appendix -2.

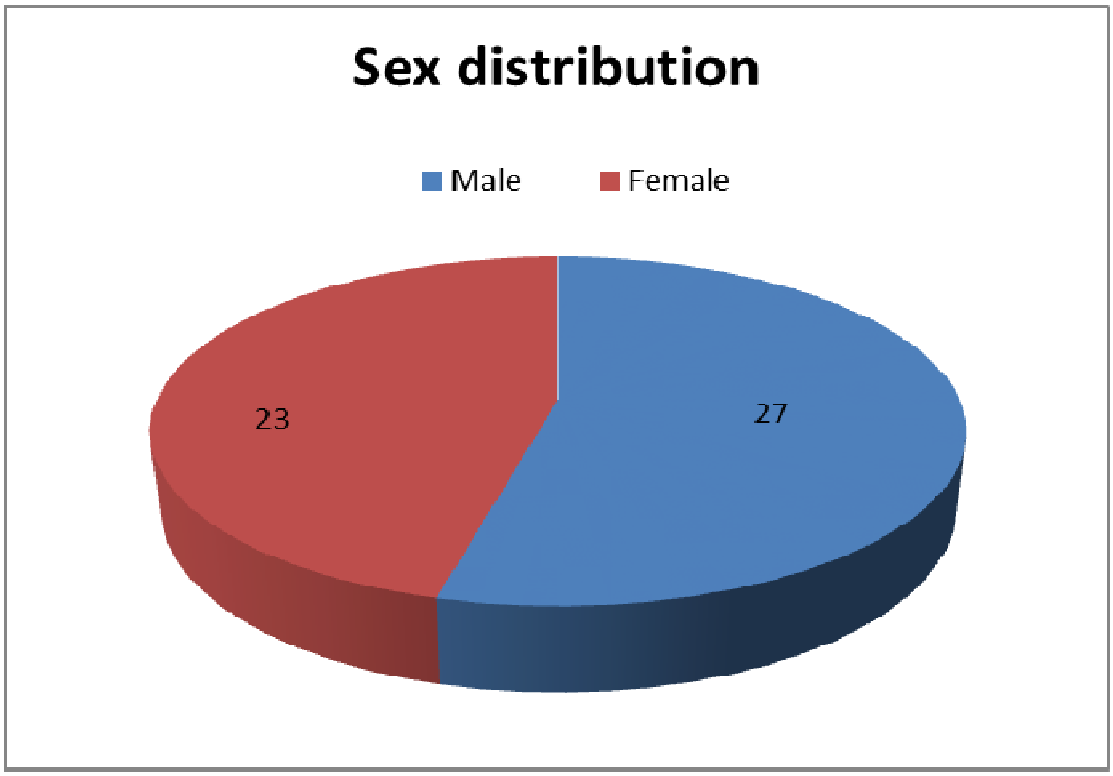
AGE DISTRIBUTION

About 14 patients were below 10 years, 13 patients were seen from 11 to 20 years, 9 patients were seen from 21 to 30 years, 10 patients from 31 to 40 years, 3 patients from 41 to 50 years and 1 patient from 51 to 60 years.



SEX DISTRIBUTION

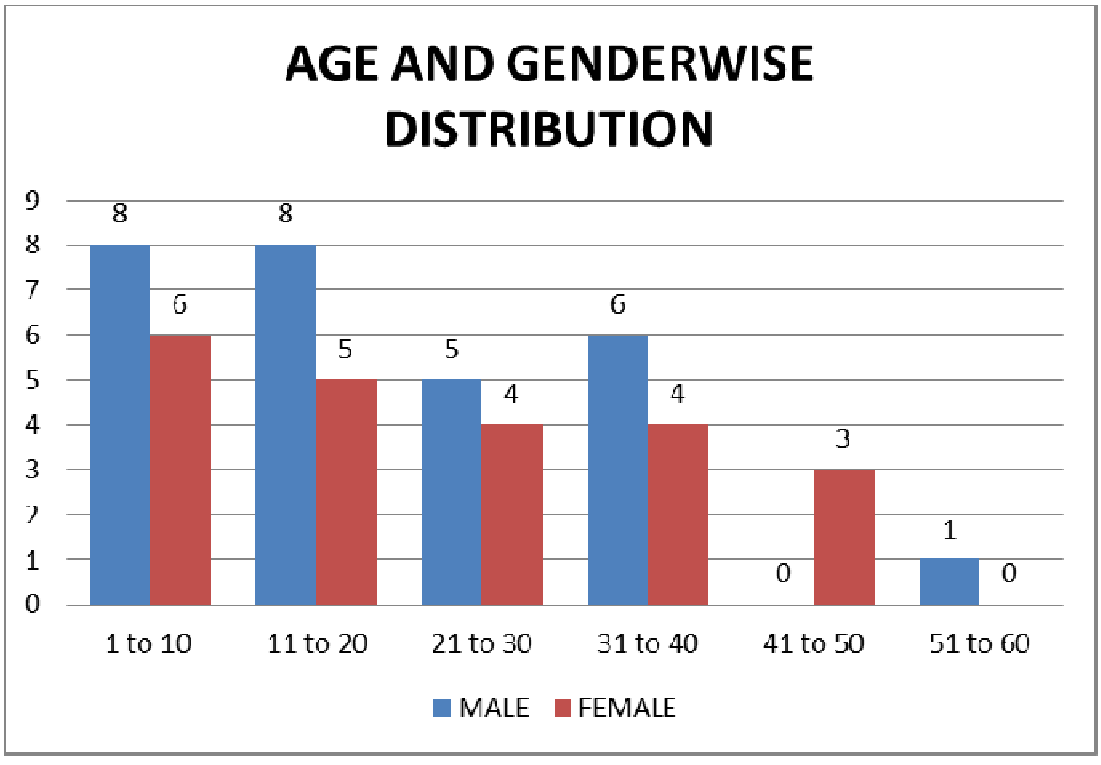
Out of 50 patients, about 27 patients were males and 23 patients were females.



DISTRIBUTION OF AGE ACCORDING TO GENDER ;

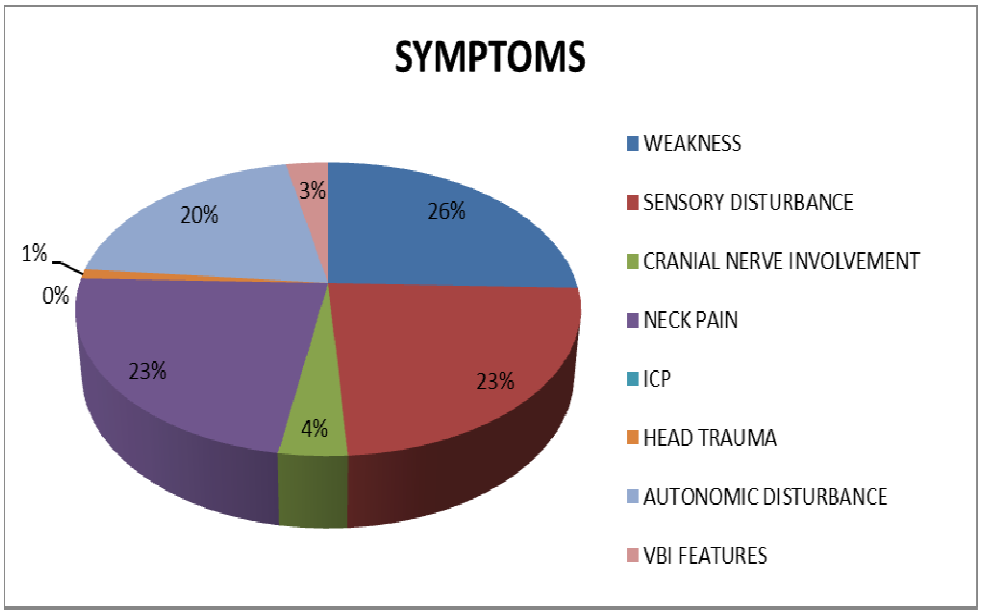
Out of 14 patients under 10 years, 8 were males & 6 were females. Under 11 to 20 years of age, about 8 patients were males & 5 patients were females, out of 13 patients. About 5 patients were males & 4 patients were under 21 to 30 years category,out of 9 patients.

Out of 10 patients under 31 to 40 years category, about 6 were males & 4 were females.All the three affected patients were females under41 to 50 years category and the only affected patient is a male under 51 to 60 years category.



DISTRIBUTION OF SYMPTOMS

About 48 patients had presented with motor weakness in the form of ascending type of spastic quadriparesis ,about 44 patients presented with sensory disturbances involving the anterolateral spinothalamic tract & Posterior column, 43 patients presented with neck pain (22 had localised Bony pain & 9 had Suboccipital Headache), about 8 patients had recent significant Head & Neck Trauma, about 38 patients presented with Autonomic disturbances (Isolated Bladder involvement in 22 patients, combined Bladder & Bowel in 38 andpatients, erectile dysfunction in 4 patients) and about 6 patients presented with features of Vertebrobasilar insufficiency in the form of vertigo.

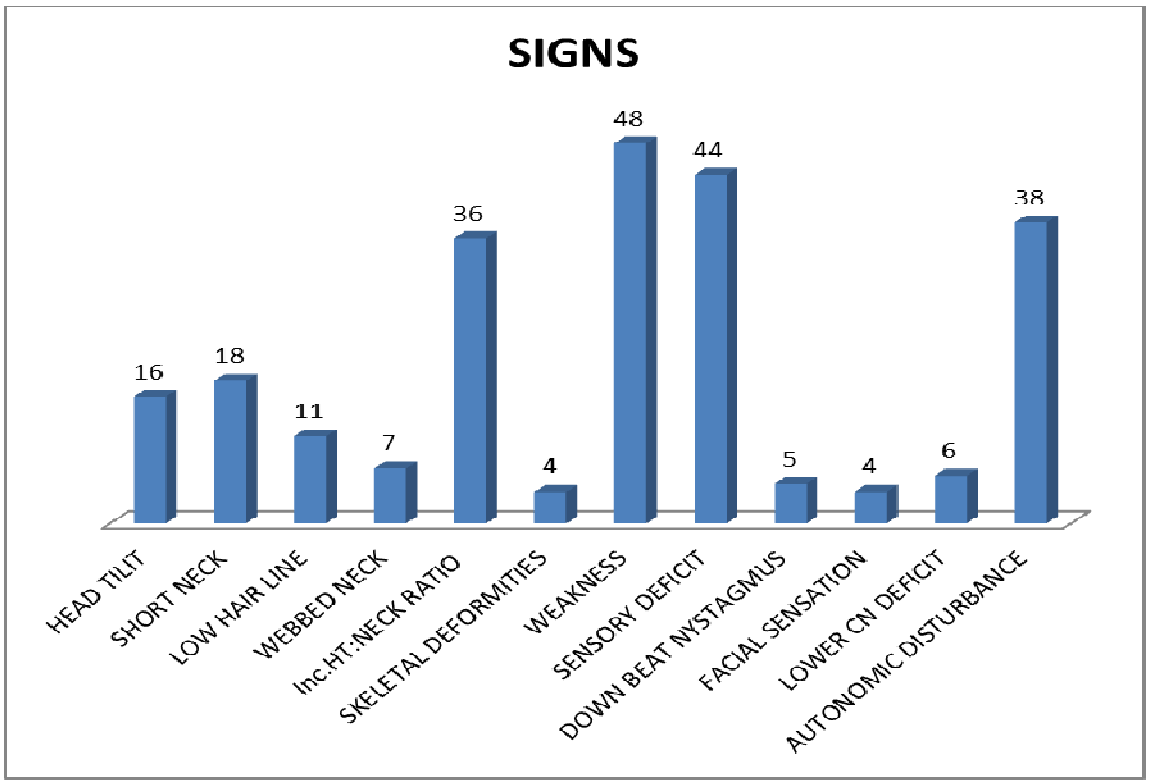


DISTRIBUTION OF CLINICAL SIGNS

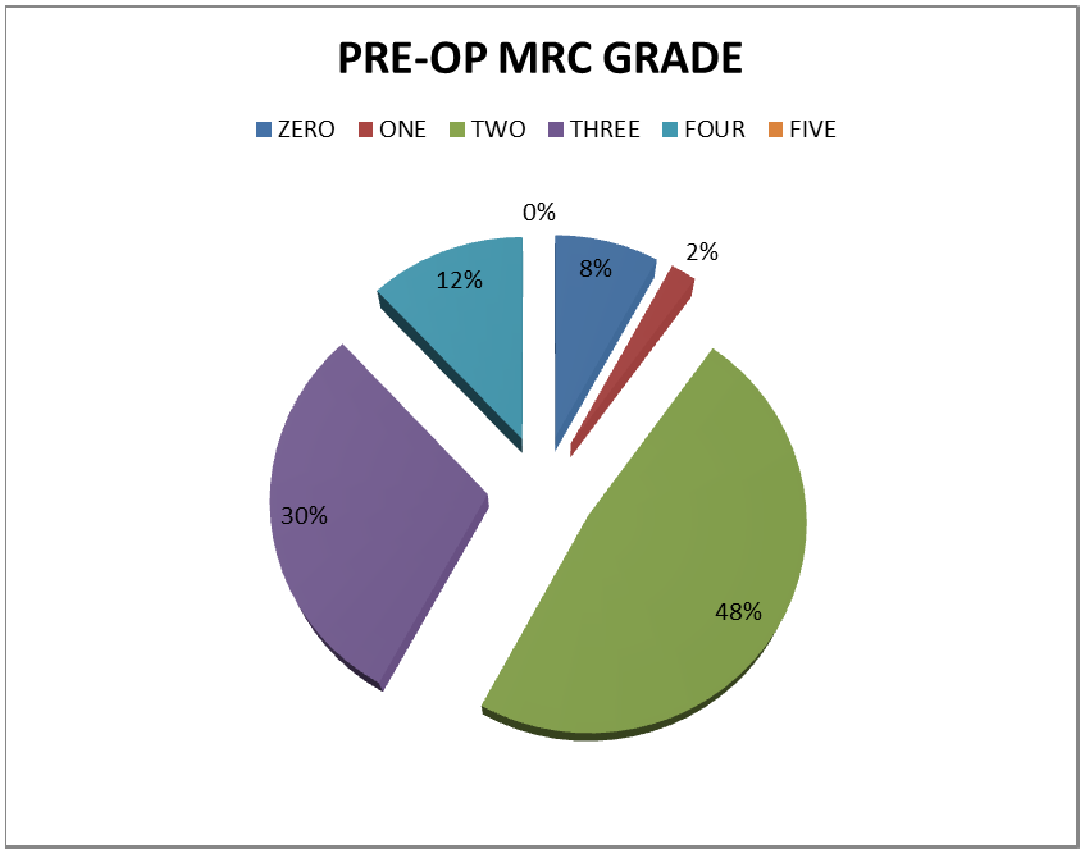
Out of 50 patients, about 16 had Head tilt, 18 had short Neck, 11 had Low hairline, 7 had Webbed neck and increased Height Neck ratio in about 29 patients.

Skeletal deformities were found in 4 patients (3 had kyphosis, 1 had scoliosis), Spastic Quadriparesis in 48 patients, sensory disturbances in 44 patients, Downbeat Nystagmus in 5 patients, Facial Sensory loss in about 4 patients, Lower Cranial nerve deficit in about 6 patients, and Autonomic disturbances in 38 patients.

DISTRIBUTION OF CLINICAL SIGNS



On examining the spinomotor system of these patients, the motor power was examined under MRC Grade. About 4 patients had Quadriplegia, one patient presented with Grade 1 power, about 24 patients had presented with Grade 2 power, about 15 patients had presented with Grade 3 power and 6 patients hadpresented with Grade 4 power.



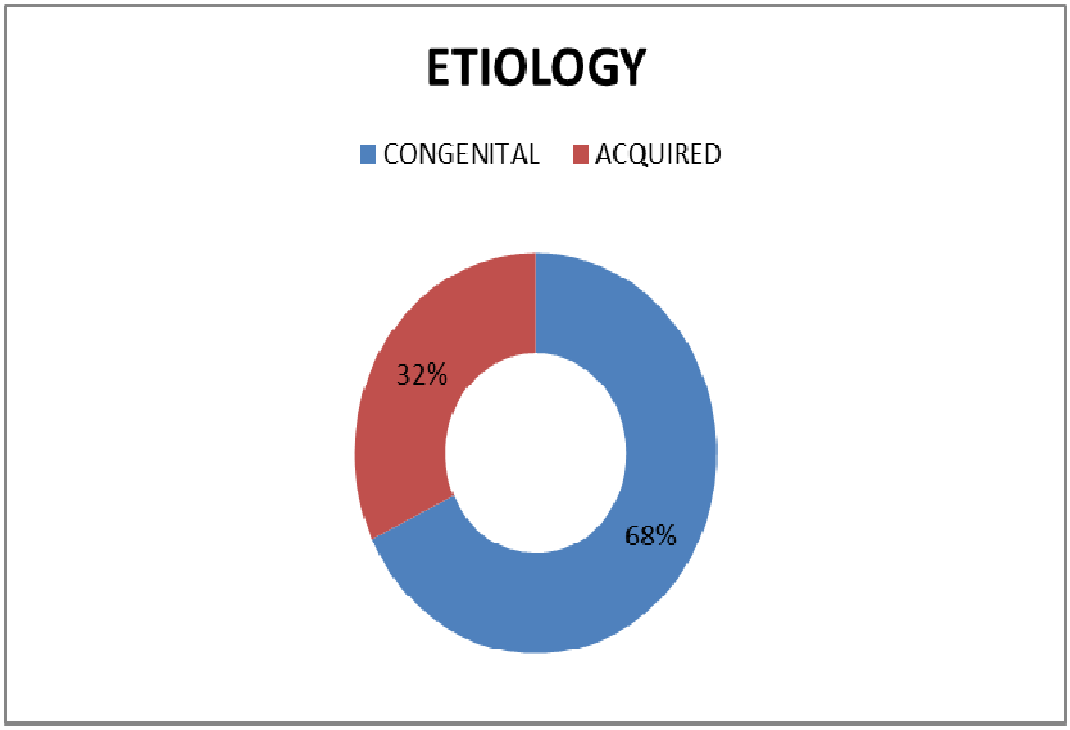
DISTRIBUTION OF ETIOLOGICAL DIAGNOSIS

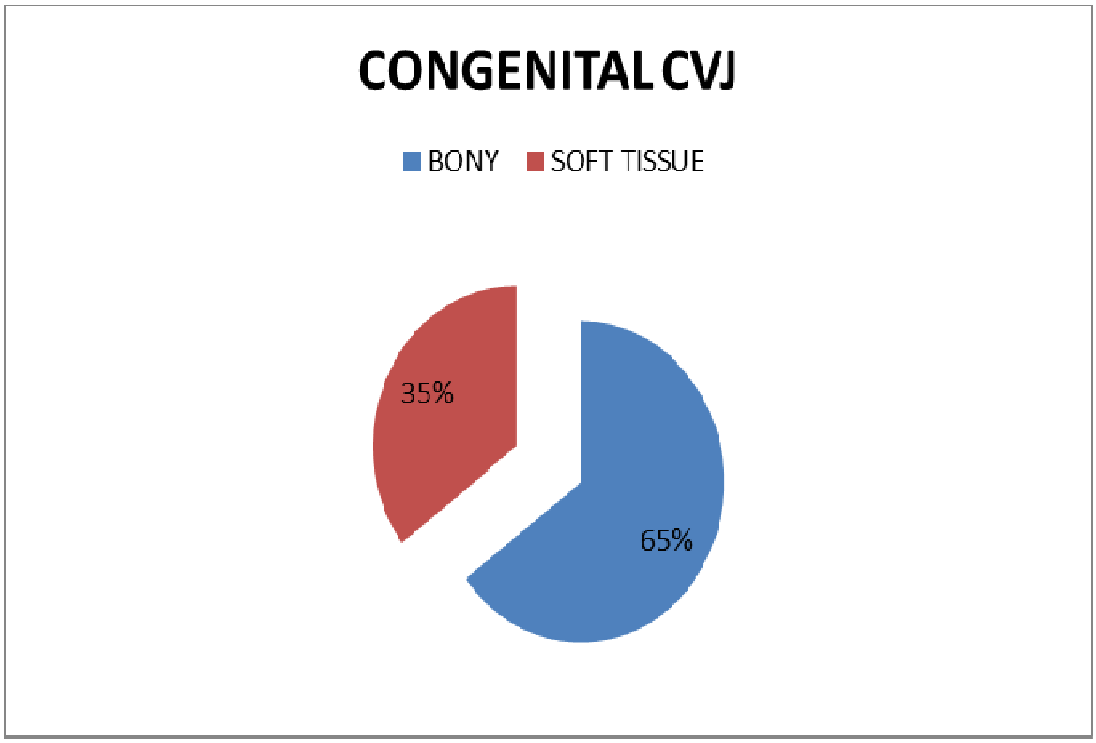
Out of 50 patients, 34 patients had congenital Anomalies and 16 patients had Acquired Anomalies. Among the 34 congenital Anomalies, 22 had Bony Anomalies & 12 had soft tissue Anomalies.Among the 22 patients, 11 had Atlantoaxial subluxation, 5 had Basilar Invagination, 3 had Platybasia, 5 had Assimilation of Atlas, 2 had defect in Anterior arch of Atlas, 1 had Os Odontoid. One patient had features of Down syndrome with Atlantoaxial subluxation.

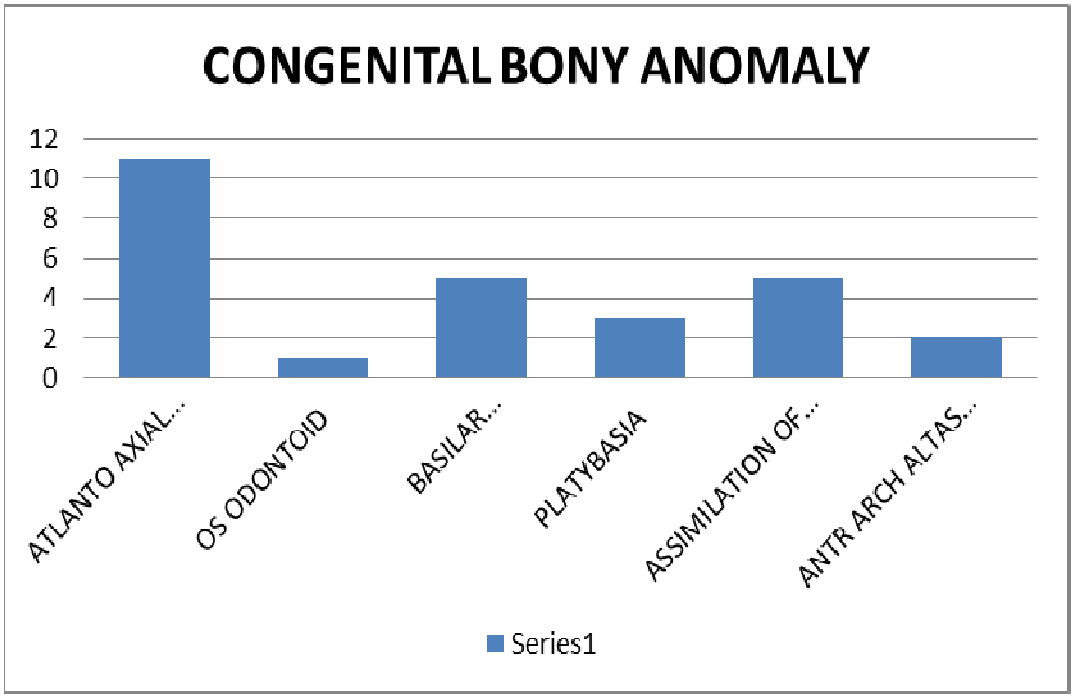
Out of 12 patients with soft tissue CVJ Anomalies, 12 had Arnold Chiari malformation.

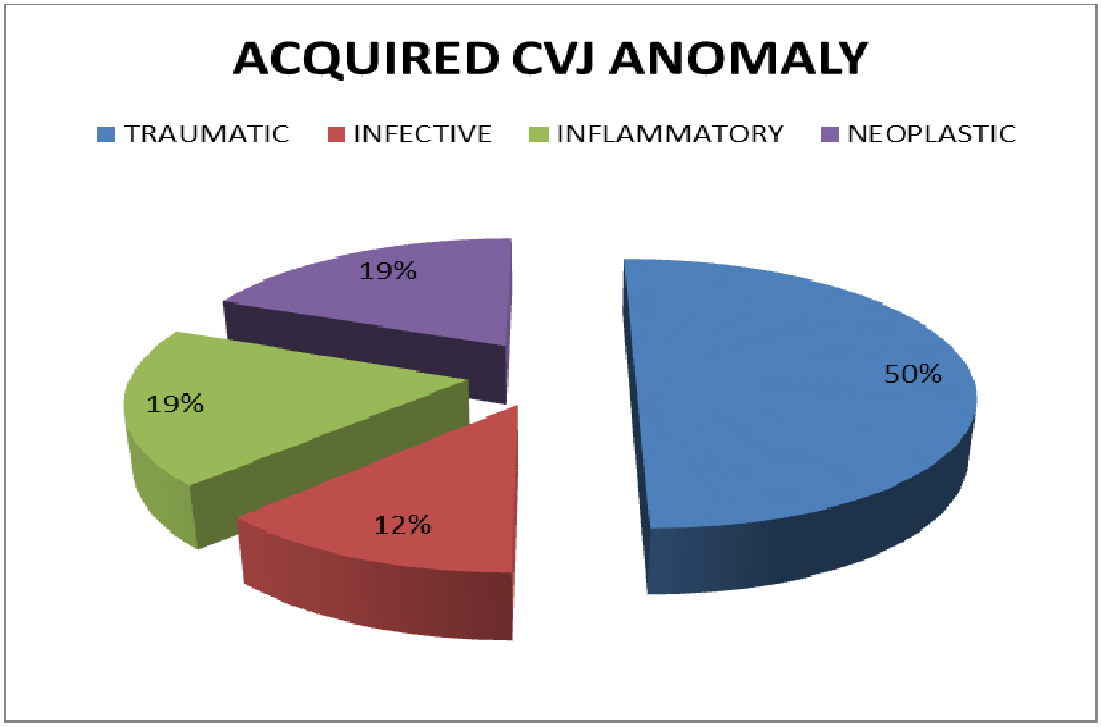
Among the Acquired CVJ Anomalies (16 patients), about 8 had developed the Anomaly following trauma, 2 had developed following Pharyngeal infection (Grisel Syndrome), 3 had developed following inflammatory causes (Tuberculosis in 2 patients & Rheumatoid Arthritis in 1 patient), 3 had developed this Anomaly following tumors (2 had Foramen magnum meningioma & 1 had Neurofibroma).

Tuberculosis CVJ Anomaly was instituted with Anti Tuberculous Therapy.


ETIOLOGY
CONGENITAL
ACQUIRED
32%
68%


CONGENITAL CVJ
BONY
SOFT TISSUE
35%
65%


CONGENITAL BONY ANOMALY
12
10
8
6
4
2
0
ATLANTO AXIAL...
OS ODONTOID
BASILAR...
PLATYBASIA
ASSIMILATION OF...
ANTR ARCH ALTAS...
Series1

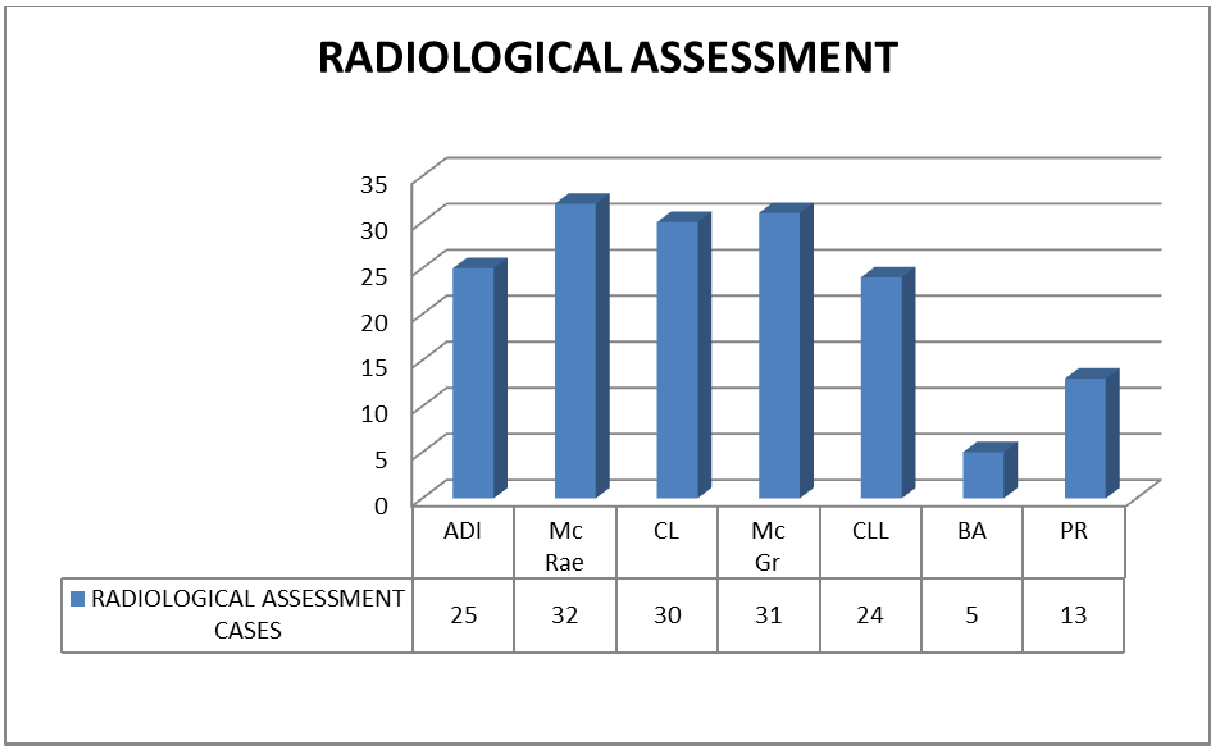

ACQUIRED CVJ ANOMALY
TRAUMATIC
INFECTIVE
INFLAMMATORY
NEOPLASTIC
19%
19%
12%
50%

DISTRIBUTION OF RADIOLOGICAL ABNORMALITIES

The Atlanto Dental Interval (ADI 3-5 mm in 13 pts, >5mm in 12 pts.) was increased in about 25 patients.

Abnormal Mc Rae's line was found in about 32 patients, Chamberlain line in 30 patients, Mc Gregor's line in 31 patients, Wackenheim Clivus Canal Line in 24patients.

AbnormalWelcher's Basal Angle >130 degrees was found in about 5 patients and the increased Power's ratio > 0.77 mm was seen in about 13 patients.



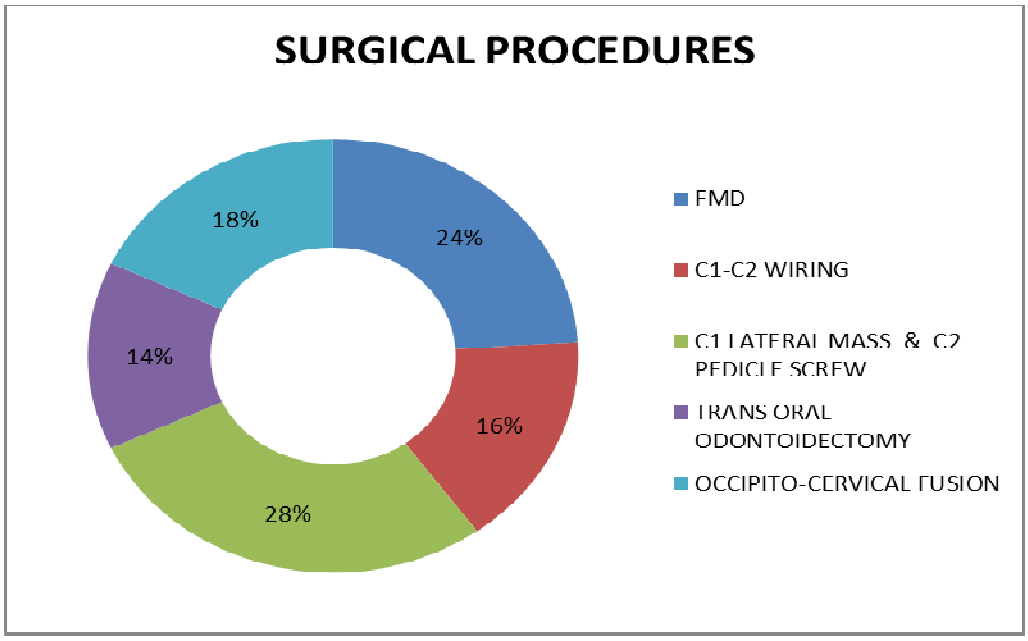
DISTRIBUTION OF VARIOUS SURGICAL TREATMENT DONE :

Out of 50 patients, about 14 patients with Atlantoaxial subluxation had underwent C1 lateral mass &C2 pedicle screw fixation. About 14 patients underwent Foramen magnum decompression for 12 patients with Arnold Chiari malformation & 2 patients with Foramen magnum tumors.

About 6 patients underwent C1 C2 wiring for Atlantoaxial Dislocation.

About 7 patients underwent TransoralOdontoidectomy(for 2 patients with Grisel syndrome, 1 with Os Odontoid, 4 patients with Basilar Invagination).

About 9 patients underwent Occipitocervical Fusion using contoured steel rod for 5 patients with Basilar Invagination & 3 patients with Platybasia, 1 with AAD.



DISTRIBUTION OF SURGICAL OUTCOME

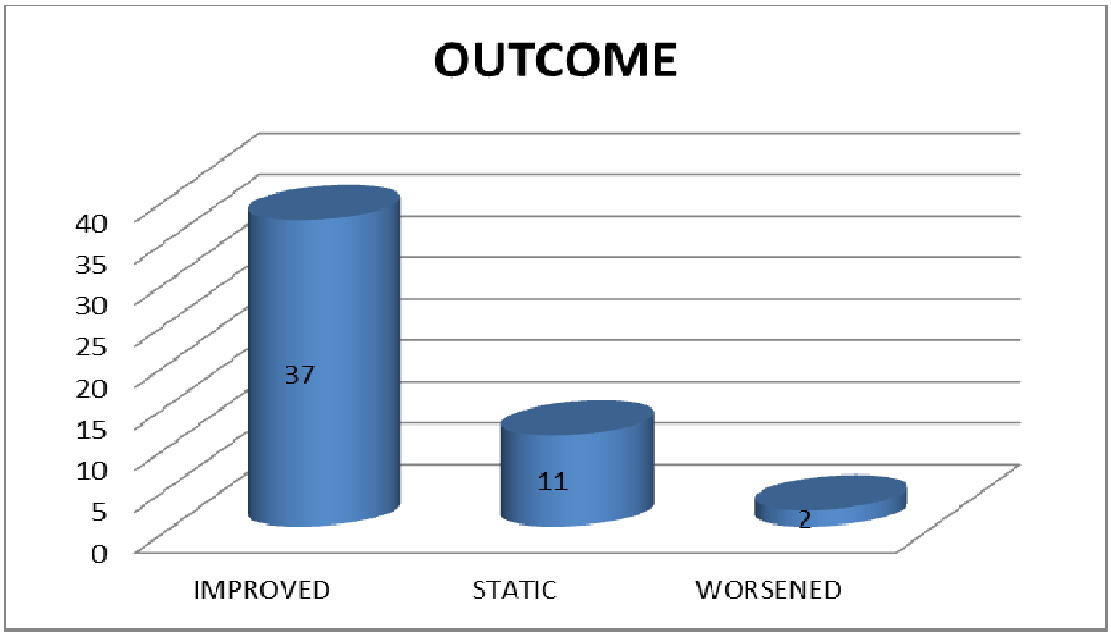
The Neurological outcome was assessed in these 50 patients by thorough clinical examination of motor power under MRC Grade.

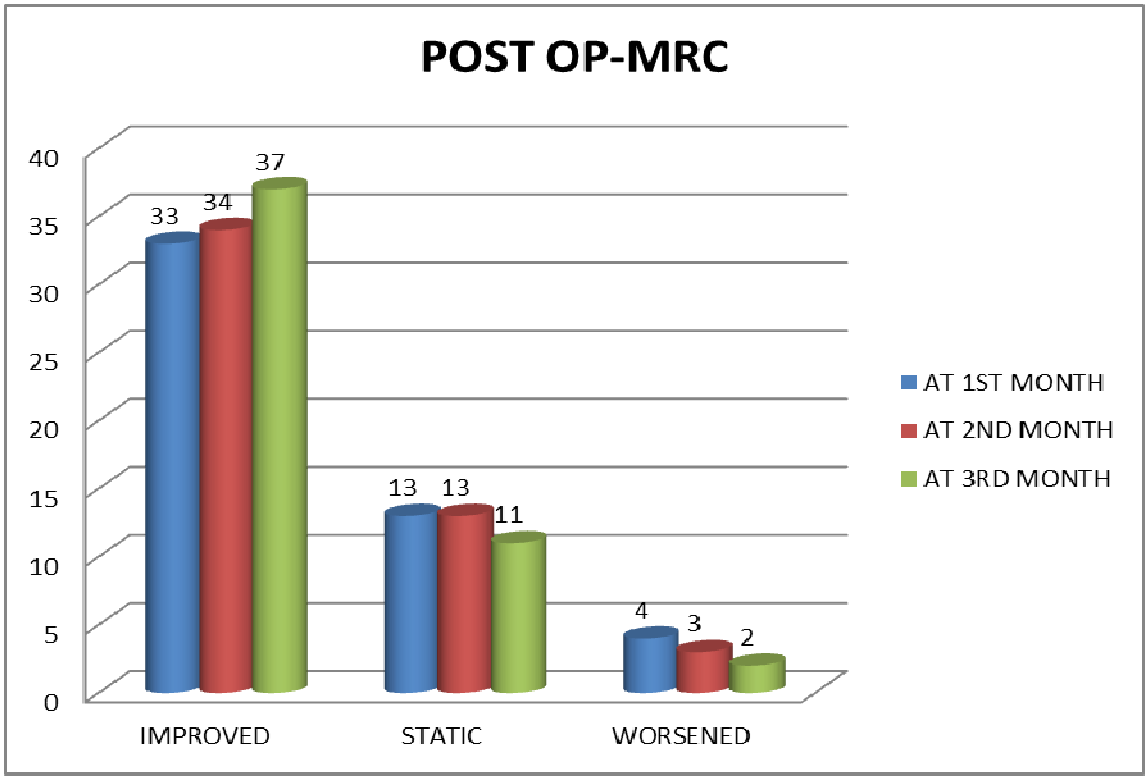
About 33 patients got improved, 13 patients had residual deficit as that of pre operative status and 4 patients got deteriorated after surgery, at 1 month follow up.

About 34 patients had improved and 13 remained static, 3 patients got worsened,at the end of 3 months follow up.

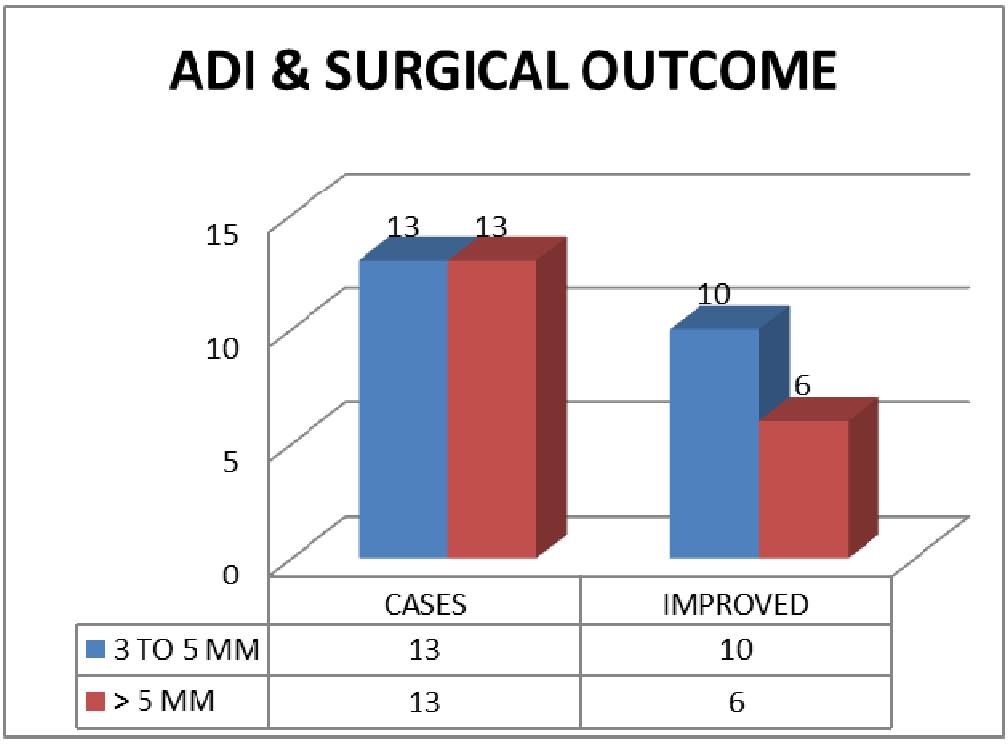
About 37 patients had improved at 6 months and 11 patients remained static, 2 patients gotdeteriorated at 6 months follow up.

All the patients had optimal reduction in thepost operative imaging.

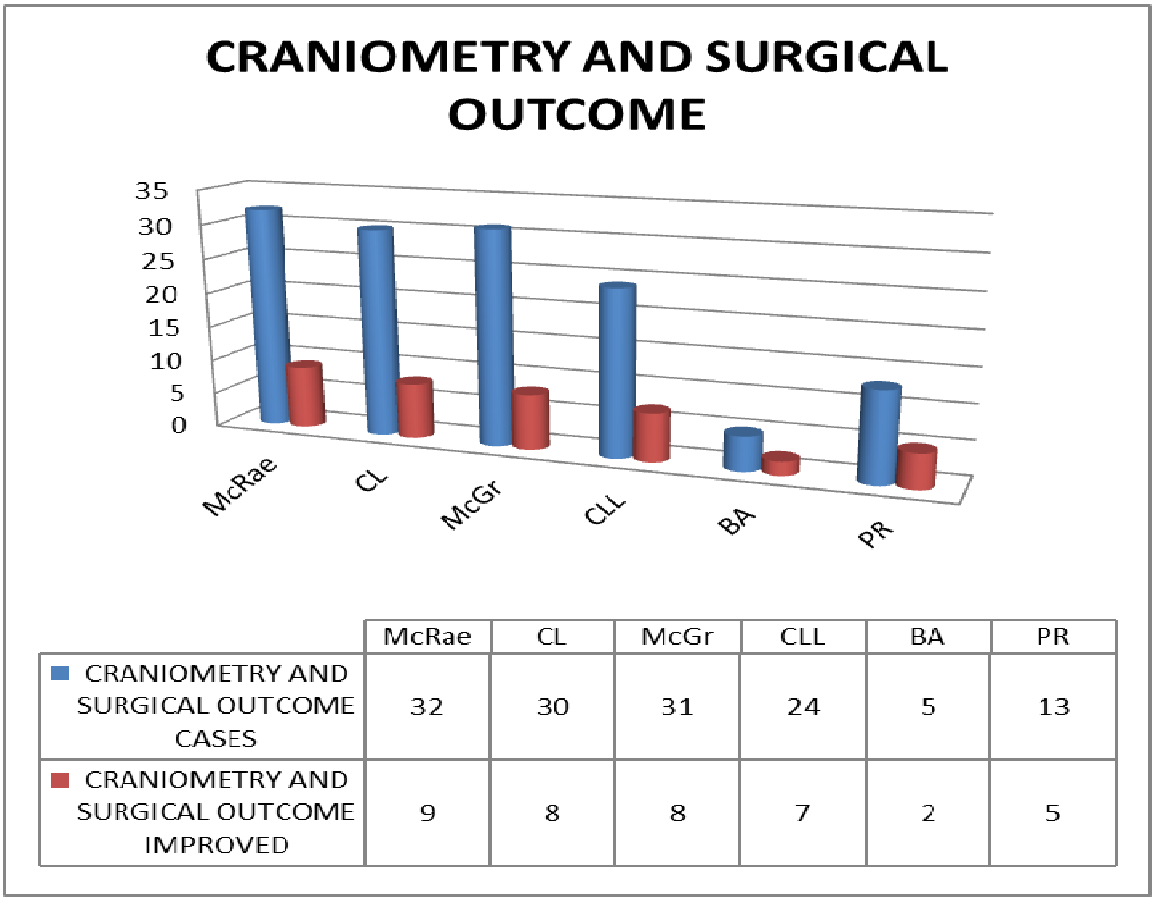

OUTCOME
40
35
30
25
20
15
10
5
0
37
11
2
IMPROVED
STATIC
WORSENED


POST OP-MRC
40
35
30
25
20
15
10
5
0
33
34
37
13
13
11
4
3
2
IMPROVED
STATIC
WORSENED
AT 1ST MONTH
AT 2ND MONTH
AT 3RD MONTH

Among the 13 patients with ADI 3-5mm, about 10 patients and among the 12 patients with ADI > 5mm, about 6 patients have showed significant postoperative improvement. On Statistical analysis by Chi-square test, the P value is 0.001 (P value<0.05) and hence it is statistically significant.



Among the 32 patients with abnormal Mc Rae's line, 9 patients have improved, 8 patients have improved out of 30 patients with abnormal Chamberlain's line, 8 patients have improved among the 31 patients with abnormal McGregor's line and 7 patients have shown improvement among the 24 patients with Wackenheim's Clivus Canal line, post operatively.



	McRae	CL	McGr	CLL	BA	PR
CRANIOMETRY AND SURGICAL OUTCOME CASES	32	30	31	24	5	13
CRANIOMETRY AND SURGICAL OUTCOME IMPROVED	9	8	8	7	2	5

STATISTICS

CRANIOMETRY& OUTCOME

Chi-squared Test for Independence

Chi-square: 1.106

Degrees of Freedom: 5

Table size: 6 rows, 2 columns.

The P value is 0.9536.

The row and column variables are not significantly associated.

Chi-square calculations are only valid when all expected

Values are greater than 1.0 and at least 80% of the expected

Values are greater than 5.

Chi-Squared Test for Trend.

Chi-squared for trend = 0.5674 (1 degree of freedom)

The P value is 0.4513.

There is not a significant linear trend among the ordered categories

defining the rows and the proportion of subjects in the left column.

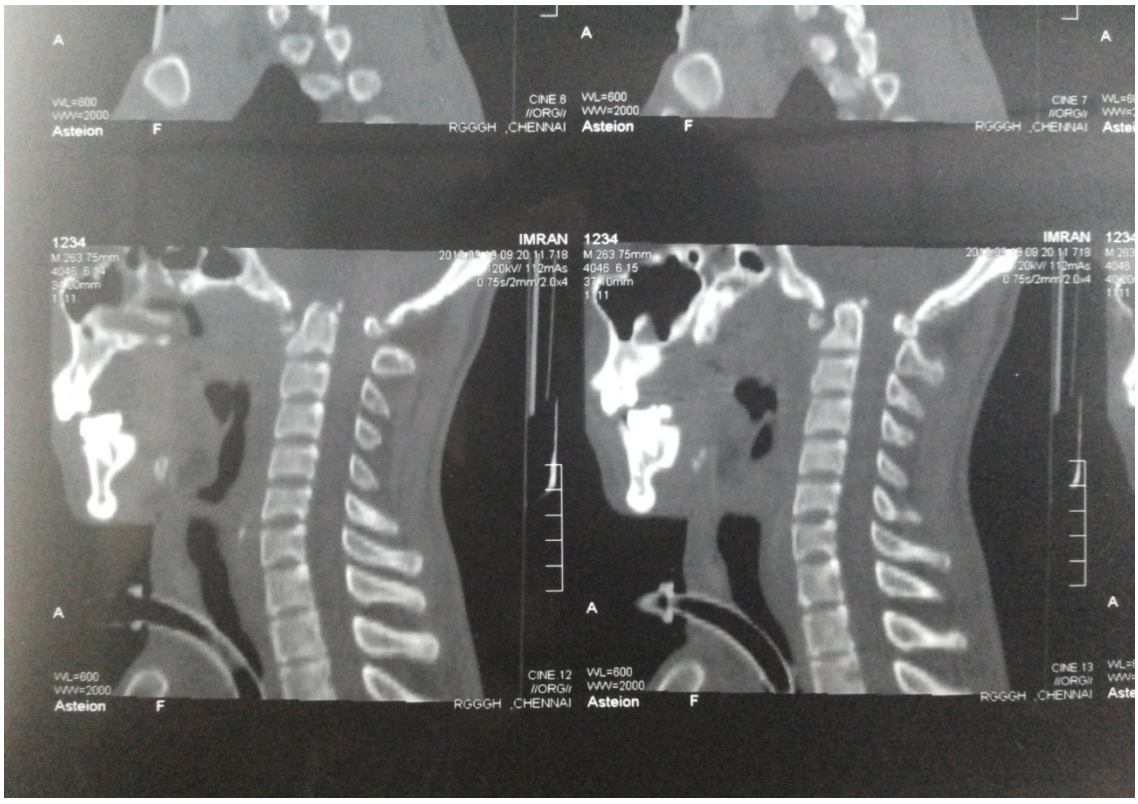
Summary of Data

Row	Total	Percent
1	32	23.70%
2	30	22.22%
3	31	22.96%
4	24	17.78%
5	5	3.70%
6	13	9.63%
Total	135	100.00%

Column	Total	Percent
NOT IMP	96	71.11%
IMPROVED	39	28.89%
Total	135	100.00%

On statistical Analysis of the Craniometric assessment and the surgical outcome, the **P value is 0.4513** (P value > 0.05) and hence it is not statistically significant.

Case No. 35,MRI CV Junction showing features of CVJ Tuberculosis

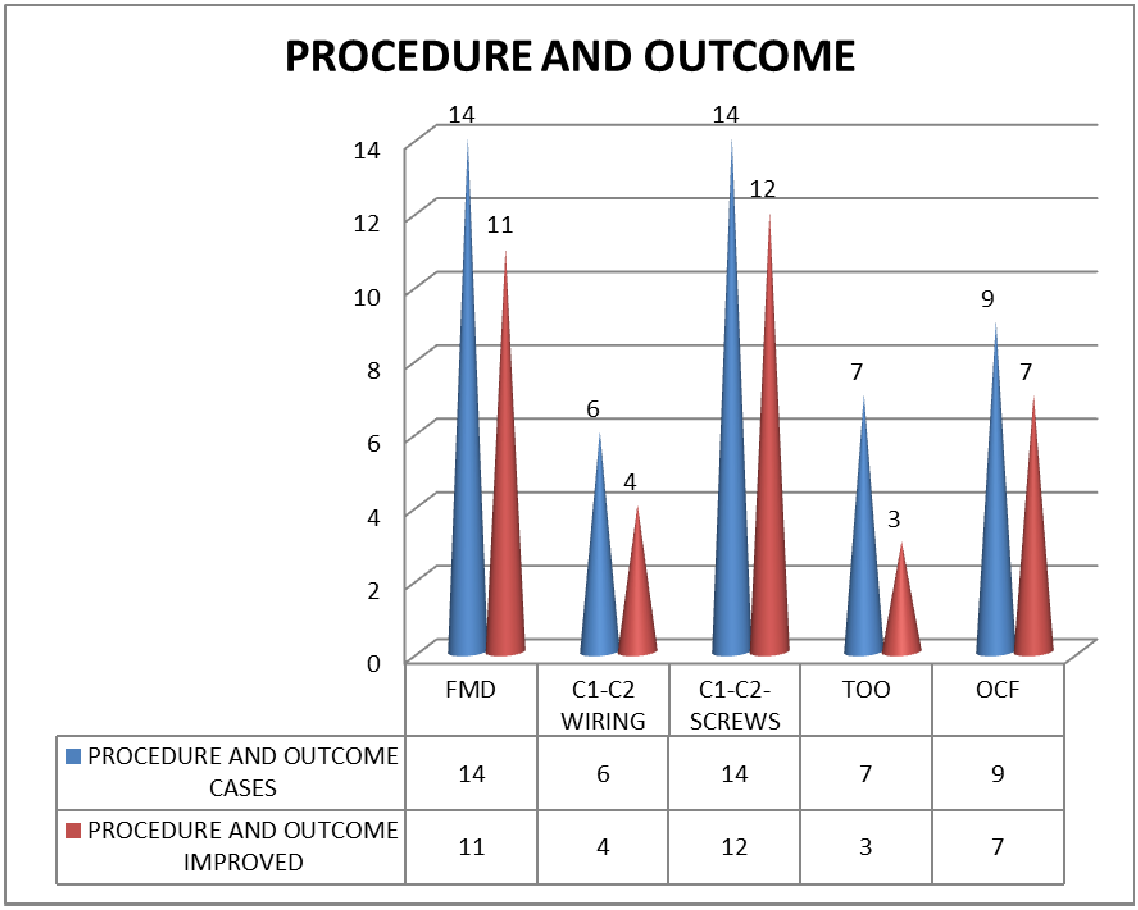


Case No. 19 showing Basilar Invagination with Assimilation of Atlas.

Post op X ray of Patient No. 18 with C1C2 wiring done.

The outcome was assessed in each surgical procedure. About 12 patients have showed significant post operative improvement in 14 patients who have undergone C1 Lateral mass & C2 pedicle screw fixation. About 11 patients have improved among the 14 patients, who haveundergone Foramen Magnum Decompression.

About 4 patients have improved among the 6 patients who have undergone C1C2 Wiring. About 3 patients have shown significant post operative outcome in the 7 patients, who have undergone TransOralOdontoidectomy and about 7 patients have improved in the 9 patients, who have undergone OccipitoCervical Fusion.


PROCEDURE AND OUTCOME
14
14
11
12
6
4
14
12
7
3
9
7
0
2
4
6
8
10
12
14
FMD
C1-C2 WIRING
C1-C2-SCREWS
TOO
OCF
PROCEDURE AND OUTCOME CASES
14
6
14
7
9
PROCEDURE AND OUTCOME IMPROVED
11
4
12
3
7

STATISTICS

SURGICAL PROCEDURES & THE OUTCOME

Chi-squared Test for Independence

Chi-square: 4.914

Degrees of Freedom: 4

Table size: 5 rows, 2 columns.

The P value is 0.2963.

The row and column variables are not significantly associated.

Chi-Squared Test for Trend.

Chi-squared for trend = 0.2753 (1 degree of freedom)

The P value is 0.5998.

There is not a significant linear trend among the ordered categories defining the rows and the proportion of subjects in the left column.

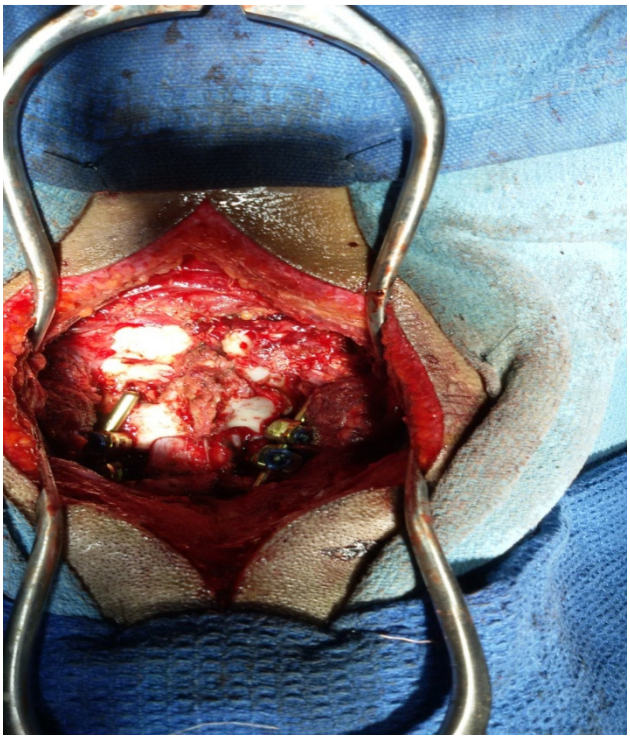
Summary of Data

Row	Total	Percent
1	14	28.00%
2	6	12.00%
3	14	28.00%
4	7	14.00%
5	9	18.00%
Total	50	100.00%

Column	Total	Percent
NOT IMP	13	26.00%
IMPROVED	37	74.00%
Total	50	100.00%

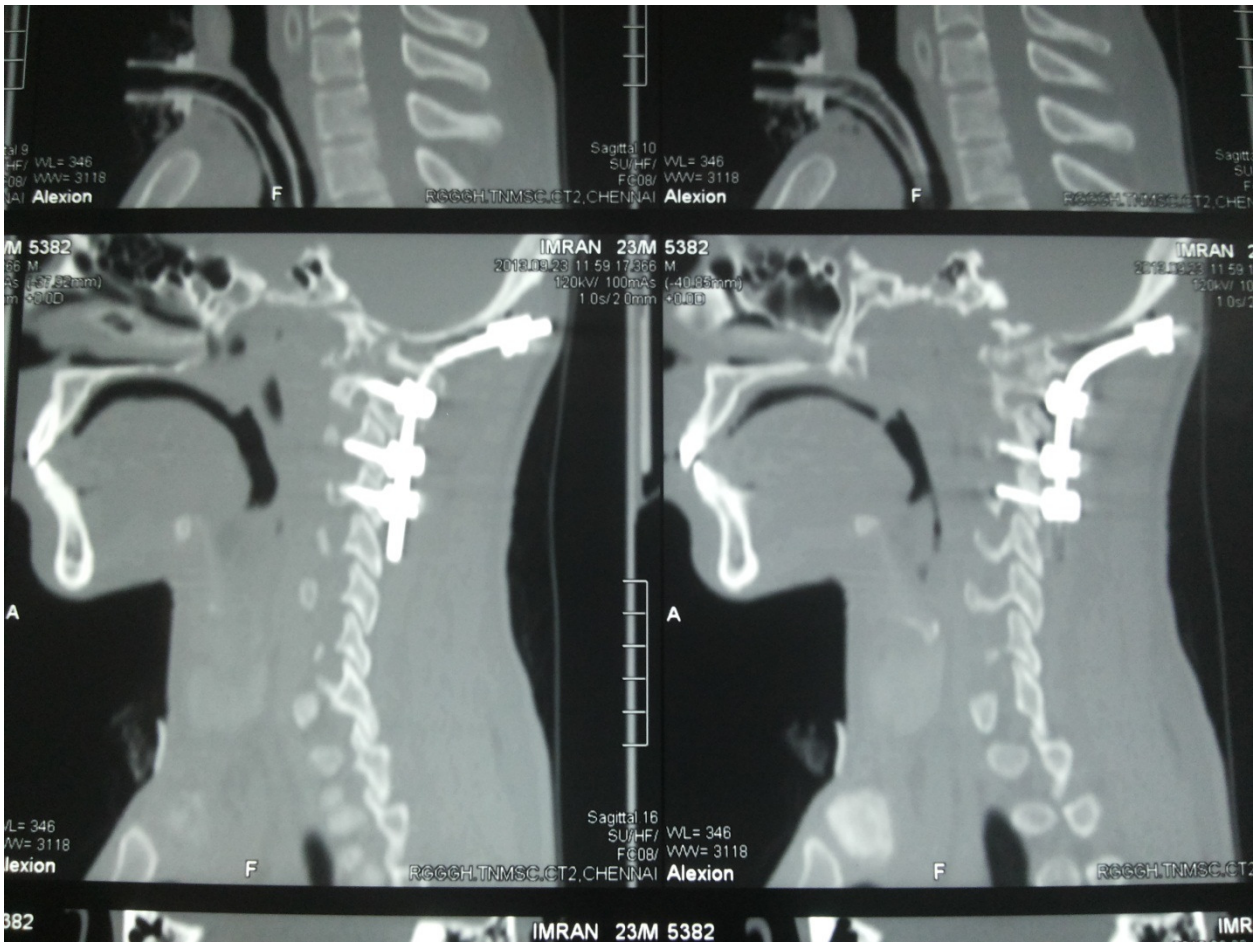
On Statistical Analysis of the different surgical procedures and the outcome, the P value is 0.5998 (P value > 0.05) and hence it is not statististically significant.

Intra op picture of C1 lateral mass & C2 pedicle screw fixation in patient No.33



Post op picture of case No.33, C1 Lateral mass C2 pedicle screw fixation done

Post op CT showing Occipitocervical Fusion in case No.44



DISCUSSION

The CV Junction Anomalies are more common in young adults (28%). The incidence is almost equal in both sexes, with slight male (54%) predominance.

The majority of CVJ Anomalies are of congenital variety (68%). Among the congenital Anomalies, the Bony anomalies (44%) are common than the soft tissue Anomalies (24%). The Alanto axial Dislocation (22%) is the most common congenital Bony Anomaly and the Arnold Chiari malformation (24%) is the most common soft tissue Anomaly. Among the Acquired CV Junction Bony Anomalies, trauma (16%) is the most common etiological factor.

These results are well comparable to the studies done worldwide, as described in the literature.

Inspite of the varied clinical presentations, motor weakness (96%), followed by sensory disturbances (80%) are the most common clinical features. Pre operative MRC Grading of motor power implies the post operative prognostic outcome.

Dynamic X ray cervical spine,Craniometric Assessment in X ray cervical spine,CT scan & MRI cervical spine are the important tool to diagnose and to tailor the surgical management. About 50% of patients had

increased Atlantodental Interval. The ADI in the preoperative imaging is well correlated with the final surgical outcome and showed statistical significance.

The Craniometric assessment showed abnormal Mc Rae's line in about 64%,Chamberlain line in 60%, McGregor's line in 62%, Wackenheim Clivus Canal Line in 48% of patients.

The increased Basal Angle was seen in about 10% and about 26% showed abnormal Power's Ratio.

Even though, the different surgeries are indicated, C1 Lateral Mass & C2 pedicle screw fixation (28%) is commonly done for Bony Anomalies and Foramen Magnum Decompression (24%) is commonly done for Soft tissue Anomalies.

About 77% of patients with abnormal ADI 3-5mm, showed significant post operative Outcome and 50% of patients with abnormal ADI >5mm have improved postoperatively. The ADI is statistically significant and well correlated with the post operative outcome. It is the most important and reliable pre operative marker in predicting the prognostic outcome.

About 28% of patients had improved postoperatively withabnormal McRae's lines, 26% of patients had improved with abnormal Chamberlain lines, 25% of patients have improved with abnormal McGregor lines.

About 29% of patients had improved with abnormal Wackenheim Clivus canal line. About 40% of patients had improved with increased Basal Angle and about 39% of patients had improved with increased Power's ratio.

The Craniometric lines are not well correlated with the final surgical outcome and it is not statistically significant.

The Craniometric lines are very arbitrary and poorly correlated with the post operative prognostic outcome.

Inspite of different surgical procedures, C1 Lateral mass & C2 Pedicle screw fixation is gaining popularity nowadays. About 86% of patients had improved after this surgery and the improvement is very less (43%) in the TransOralOdontoidectomy.

About 79% of patients had shown improvement following Foramen magnum Decompression and about 67% of patients had improved following C1C2 Wiring.

About 78% of patients had improved following Occipitocervical Fusion using contoured rod.Inspite of different surgical techniques described, no particular technique is bettercorrelated with the post operative outcome and it is statistically insignificant. Hence, the each surgical procedure may be indicated depending on the case.This is comparable to the studies done

worldwide, where there are no definite indications for each surgical technique and no consensusis described in the literature.

Many patients have improved neurologically (54%), after surgery at the end of 6 months follow up.

Since the sample size is very small, the study with large sample size is needed to obtain still more better results.

CONCLUSION

1) Among all the Craniovertebral Junction Anomalies, Bony Anomalies are more common than soft tissue Anomalies. It is more common in young adults. It is almost equal in both the sexes with slight male predominance.

2) Congenital CVJ Anomalies are more frequent than Acquired Anomalies.

3) Atlanto Dental Interval is the single important & reliable marker to assess the prognostic outcome.

4) Craniometric assessment is very arbitrary and not correlated with the final surgical outcome.

5) Inspite of different surgical procedures, each surgical technique is tailored according tothe patient.

6) Early surgery carries good prognostic outcome especially in patients with better motor power.

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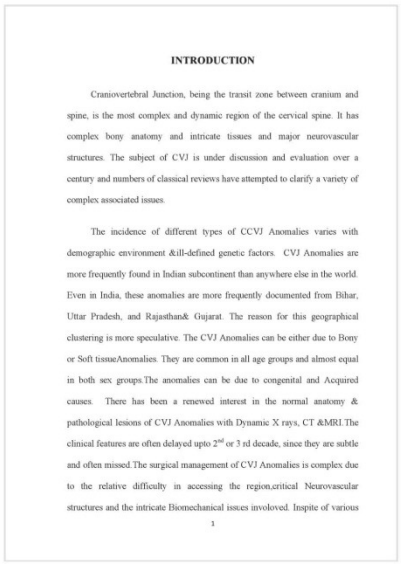
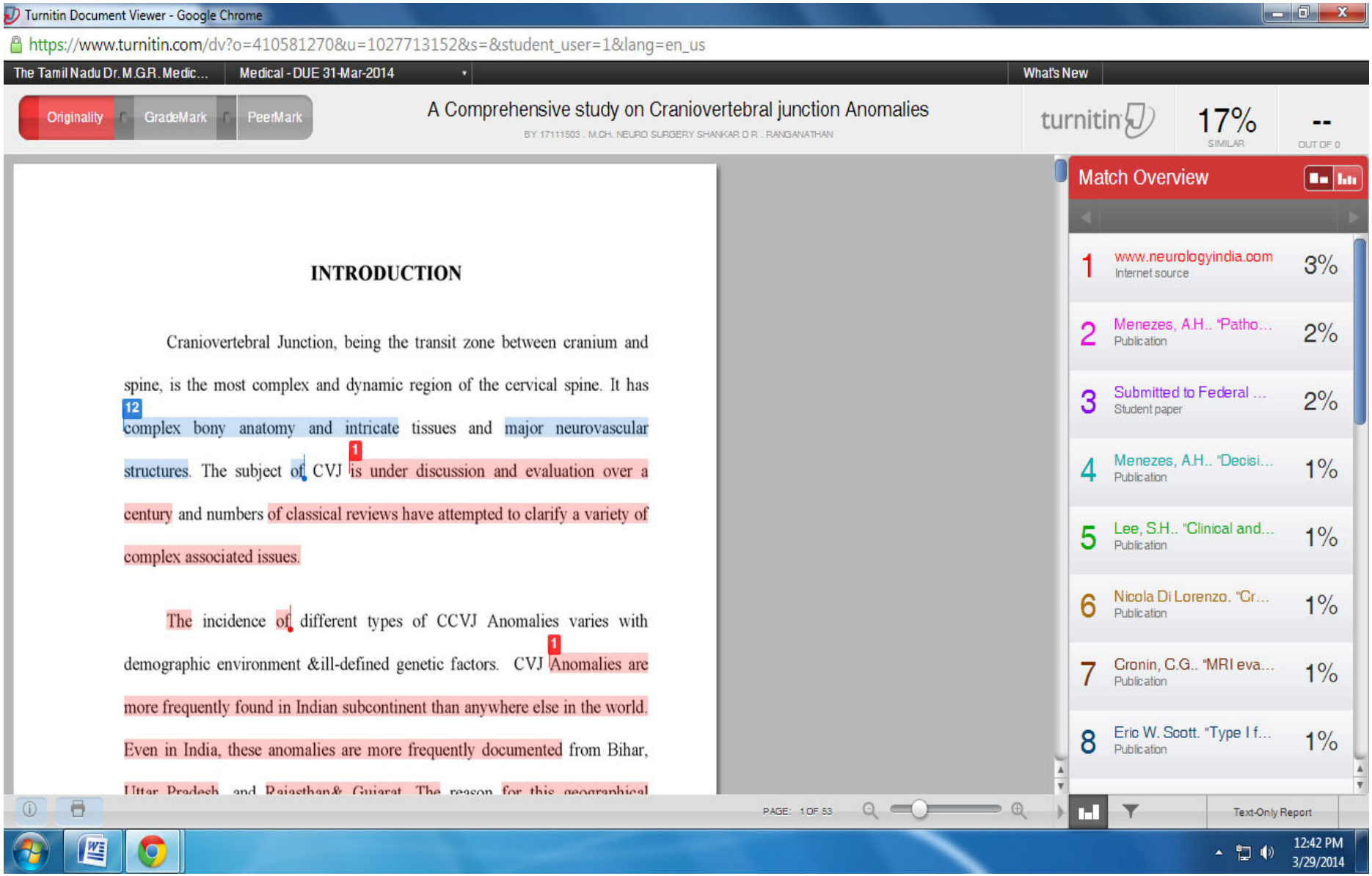


INTRODUCTION

Craniovertebral Junction, being the transit zone between cranium and spine, is the most complex and dynamic region of the cervical spine. It has complex bony anatomy and intricate tissues and major neurovascular structures. The subject of CVJ is under discussion and evaluation over a century and numbers of classical reviews have attempted to clarify a variety of complex associated issues.

The incidence of different types of CCVJ Anomalies varies with demographic environment &ill-defined genetic factors. CVJ Anomalies are more frequently found in Indian subcontinent than anywhere else in the world. Even in India, these anomalies are more frequently documented from Bihar, Uttar Pradesh, and Rajasthan& Gujarat. The reason for this geographical clustering is more speculative. The CVJ Anomalies can be either due to Bony or Soft tissueAnomalies. They are common in all age groups and almost equal in both sex groups.The anomalies can be due to congenital and Acquired causes. There has been a renewed interest in the normal anatomy & pathological lesions of CVJ Anomalies with Dynamic X rays, CT &MRI.The clinical features are often delayed upto 2nd or 3 rd decade, since they are subtle and often missed.The surgical management of CVJ Anomalies is complex due to the relative difficulty in accessing the region,critical Neurovascular structures and the intricate Biomechanical issues involoved. Inspite of various

1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INTRODUCTION
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INSTITUTE OF NEUROLOGY

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PROFORMA

Serial No :

Name: MIN No :
Age : M / F I.P. No. Occupation :

Chief Complaints :

Duration of Complaints :

History

History of Spinomotor system involvement :

History of sensory disturbances :

History of Visual disturbances :

History of Lower cranial Nerves Involvement :

History of Neck pain :

History suggestive of increased ICT :

History of head \ neck trauma :

History of sphincter disturbances :

History of miscellaneous complaints :
(giddiness / syncope / LOC)

General Examination

Head tilt (Torticollis) :

Short neck :

Low hairline :

Webbed neck :

Height / Neck Ratio :

Skeletal dysplasia :

Spinal deformities :

Motor Weakness AL`54
Quadriparesis \ paraparesis \ monoparesis :
MRC Grade -
Renawat classification -
Frenkel Grade -
Nurick's Grade. -

Nystagmus – downbeat \ gaze evoked :

Facial Sensation :

Lower Cranial Nerves :

Bladder \ Bowel disturbances :

Investigations

X Ray Cervical Spine – Dynamic X Rays:

AP \ Lat \ Open Mouth views :

Mc Rae's line :

Mc Gregor line :

Chamberlain line :

Wackenheim clivus canal line :

Atrlanto Dental Interval (ADI) :

Powers ratio :

Welcher's Basal Angle :

CT Cervical Spine :

MRI Cervical Spine :

Etiological Diagnosis:

1. Congenital : Yes / No
2. Acquired : Yes / No

a) Trauma : Yes / No
b) Infection : Yes / No
c) Inflammation : Yes / No
d) Neoplastic : Yes / No
e) Idiopathic: Yes / No

Final Diagnosis

Congenital \ Acquired :

Type of CVJ Anomaly :

Management

Surgery done :

Intraop complication :

Outcome

Neurological improvement :

Any residual deficit :

Death - sequence of events – Cause :

Follow Up

One month – improved \ deteriorated :

Six month - improved \ deteriorated :

MASTER CHART

S. NO.		SYMPTOMS								SIGNS												RADIOLOGICAL INV.									ETIOLOGY											SURGERY DONE				
	A/s	SMS	S	CN	P	ICP	Tr	Aut.	VBI	Ti	SN	LH	WN	H/N	SK	W	MRC	SK	DN	F	LCN	AD	ADI	MR	MG	CL	WCC	BA	PR	CT	MRI	AAD	OO	BI	PB	AA	AD	AC	Tr	Inf.	Infl.	Neo.	FMD	Wiring	C1C2 Fixn.	TOO	OCF
1	12/M	1	0	0	1	0	0	0	0	0	0	0	0	1	0	1	0	0	0	0	0	0	1	1	1	1	0	0	0	1	1	0	0	0	0	0	0	0	0	1	0	0	1	0	0	0
2	7/F	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	1	0	0	0	0	1	0	0	0
3	44/M	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	0	0	0	1	0	1	0	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	1
4	52/F	1	1	0	1	0	0	0	0	1	1	0	0	1	0	1		0	0	0	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	1	1
5	28/M	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	2	0	0	0	0	1	0	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	1
6	33/F	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	3	0	0	0	0	1	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	1	0	0
7	9/M	1	1	0	1	0	0	1	0	1	1	1	1	1	0	1	4	0	0	0	0	1	0	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	0
8	15/F	1	1	0	1	0	0	0	0	0	0	0	0	1	0	1	2	0	0	0	0	0	1	1	1	1	1	0	1	1	1	1	0	0	0	0	0	0	0	0	0	0	1	0	0	0
9	37/M	1	1	0	1	0	1	0	0	0	0	0	0	1	0	1	2	0	0	0	0	1	1	1	1	1	1	0	1	1	1	0	0	0	0	0	0	1	0	0	0	0	0	0	0	1
10	7/F	1	1	0	0	0	0	1	0	1	1	1	1	1	0	1	3	0	1	0	0	1	1	1	1	1	1	0	1	1	1	0	0	0	0	0	0	0	1	0	0	0	0	1	0	0
11	19/M	1	1	0	1	0	0	0	0	0	0	0	0	1	0	1	2	1	0	1	0	0	1	1	1	1	1	1	1	1	1	0	0	1	0	1	0	0	0	0	0	0	0	0	0	1
12	31/F	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	3	0	0	0	0	0	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	1	0	0
13	24/M	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	2	0	0	0	0	1	0	1	1	1	1	0	1	1	1	0	0	0	1	0	0	0	0	0	0	0	0	1	0	0
14	18/F	1	1	0	1	0	0	1	0	0	0	0	0	0	0	1	3	0	0	1	0	1	1	1	1	1	1	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	0	0	1
15	9/M	1	1	1	1	0	0	0	0	1	1	1	1	1	1	1	2	0	1	0	0	1	1	1	1	1	1	0	1	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0
16	39/F	1	1	0	1	0	1	0	0	0	0	0	0	0	0	1	2	0	0	0	0	1	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0
17	16/M	1	1	0	1	0	0	1	1	0	1	0	0	1	0	1	2	0	0	0	0	0	0	0	0	0	1	1	1	1	1	0	0	0	1	0	0	0	0	0	0	0	0	0	0	0
18	34/F	1	1	0	0	0	0	0	0	0	0	0	0	1	0	1	3	0	0	0	0	1	0	0	0	0	0	1	1	1	1	0	0	1	0	1	0	0	0	0	0	0	0	1	0	0
19	17/M	1	1	0	1	0	0	0	0	1	1	0	0	1	1	1	3	1	0	1	0	1	1	1	0	0	0	0	1	1	1	0	0	0	0	0	1	0	0	0	0	0	0	0	0	0
20	33/F	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	2	0	0	1	0	1	1	1	1	1	1	0	0	1	1	0	0	0	0	0	0	0	0	1	0	0	0	0	1	1
21	16/M	1	0	1	1	0	1	0	0	1	1	1	0	1	1	1	2	0	0	0	0	1	1	0	1	0	0	0	0	1	1	0	1	0	0	0	0	0	0	0	0	0	0	0	1	1
22	37/F	1	1	0	1	0	0	0	1	0	0	0	0	1	0	1	3	0	0	0	0	1	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	1	0	0
23	8/M	1	1	0	1	0	0	0	0	1	1	0	1	1	1	1	3	1	1	0	0	1	1	1	1	1	1	0	0	0	1	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0
24	9/F	1	1	0	1	0	0	0	0	1	1	1	1	1	0	1	2	0	0	0	0	1	1	0	0	0	0	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	0	0	0
25	27/M	0	1	0	0	0	0	1	0	0	0	0	0	1	0	0	2	0	0	0	0	1	0	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	0
26	36/F	1	1	1	1	0	1	0	0	0	0	0	0	0	0	1	2	0	0	1	0	0	1	1	1	0	0	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	0	1	1
27	22/M	1	1	1	1	0	0	1	0	0	0	0	0	1	0	1	3	0	0	0	0	1	1	1	0	0	0	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0
28	33/F	1	0	0	1	0	0	0	1	0	0	0	0	1	0	1	2	0	0	0	0	1	0	0	0	0	1	1	1	1	1	0	0	0	1	0	0	0	0	0	0	0	0	1	0	0
29	29/M	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	3	0	0	1	0	1	0	1	1	1	1	0	0	1	0	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0
30	26/F	1	1	0	1	0	0	0	0	0	1	0	0	0	0	1	2	0	0	0	0	1	1	1	0	0	1	0	0	1	1	0	0	0	0	0	1	0	0	0	0	0	0	1	0	1
31	18/M	1	1	0	1	0	1	0	0	1	1	0	0	0	0	1	2	0	0	0	0	0	1	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	0
32	7/F	1	1	0	0	0	0	1	1	1	1	0	0	0	0	1	3	0	1	0	1	1	1	1	1	1	0	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0
33	24/M	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	3	0	1	0	1	1	1	1	1	1	0	0	0	1	1	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0
34	46/F	1	1	1	1	0	0	1	0	0	0	0	0	0	0	1	2	0	0	0	0	1	0	0	0	0	1	1	1	1	1	0	0	1	0	1	0	0	0	0	0	0	0	0	0	0
35	33/M	1	0	0	1	0	0	0	0	0	0	0	0	0	0	1	4	0	0	0	0	1	1	1	1	1	1	0	0	1	1	0	0	0	0	0	0	0	0	0	1	0	0	0	0	0
36	16/F	1	1	0	1	0	0	0	0	1	1	1	0	1	0	1	2	0	0	0	0	0	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	1	0	0
37	41/M	1	1	0	0	0	0	0	1	0	0	0	0	0	0	1	2	0	0	0	1	1	0	1	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	1	1	0	1	0	0
38	32/F	1	1	0	1	0	0	0	0	0	0	0	0	0	0	1	3	0	0	0	0	1	1	1	1	1	0	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	0	0	0
39	9/M	1	1	0	1	0	1	1	0	1	1	1	0	1	0	1	4	1	1	0	1	1	0	1	1	1	0	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	1	0	0
40	8/F	1	0	1	1	0	0	0	0	1	1	1	0	1	0	1	2	0	0	0	0	1	0	1	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	1	0	0	0	0
41	19/M	1	1	0	0	0	0	1	0	0	0	0	0	1	0	1	3	0	0	0	0	1	1	0	1	0	0	0	1	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0
42	26/F	0	1	1	1	0	0	0	0	0	0	0	0	1	0	0	2	0	0	1	0	1	0	1	1	1	0	0	0	0	1	0	0	0	0	0	0	0	0	0	1	1	0	0	0	0
43	8/M	1	1	0	1	0	1	0	0	1	1	1	1	1	0	1	2	0	0	0	0	1	1	1	0	0	0	0	0	1	1	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0
44	25/F	1	1	0	0	0	0	1	0	0	0	0	0	1	0	1	3	0	0	0	0	1	1	0	0	0	0	1	1	1	1	1	0	0	1	0	1	0	0	0	0	0	0	1	0	0
45	7/M	1	0	0	1	0	0	0	1	1	1	1	1	1	0	1	4	0	0	0	1	1	1	1	1	1	1	0	0	1	1	1	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0
46	27/F	1	1	0	1	0	1	0	0	0	0	0	0	1	0	1	2	0	0	0	0	0	1	1	0	0	0	0	0	0	0	1	0	0	0	0	0	1	0	0	0	0	0	1	0	1	0	0
47	10/M	1	1	0	1	0	0	1	0	1	1	1	0	1	0	1	2	0	0	0	1	1	0	1	0	0	0	0	0	1	1	0	0	0	0	0	0	0	0	0	0	1	1	0	0	0	0
48	6/F	0	0	0	0	1	0	0	0	1	1	1	0	1	0	0	4	0	0	0	0	2	0	1	0	1	0	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	0	1	0	0	0	0
49	22/M	1	1	1	0	1	0	0	0	8	1	1	1	0	0	1	2	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	1	0	1	0	0
50	40/M	1	1	1	0	1	0	0	0	7	1	1	1	0	0	0	4	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0	0	0	1	0	0	0	0	0	0	0

KEY WORDS TO MASTER CHART

AA	–	Assimilation of Atlas
AAD	–	Atlantoaxial Dislocation
AC	–	Arnold Chiari malformation
AD	–	Autonomic disturbances
AD	–	Defect in the Atlas arch
ADI	–	Atlantodental Interval
Aut.	–	Autonomic disturbances
BA	–	Basal Angle
BI	–	Basilar Invagination
C1C2 Fix	–	C1 Lateral mass & C2 Pedicle screw Fixation
CCL	–	Clivus canal line
CL	–	Chamberlain line
CN	–	Cranial nerve disturbances
DN	–	Downbeat Nystagmus
FMD	–	Foramen magnum Decompression
H/N	–	Increased Height Neck Ratio
ICP	–	Increased Intracranial Pressure symptoms
LCN	–	Lower Cranial Nerves
LH	–	Low Hairline
MG	–	Mc Gregor line
MR	–	Mc Rae’s line
OCF	–	Occipito cervical Fusion
OO	–	OsOdontoideum
P	–	Neck pain
PB	–	Platybasia
PR	–	Power’s ratio

SK	–	Skeletal deformity
SMS	–	Spinomotor system
SN	–	Short Neck
Ti	–	Head Tilt
TOO	–	TransoralOdontoidectomy
Tr	–	Trauma
VBI	–	Vertebrobasilar Insufficiency
W	–	Weakness
Wiring	–	C1 C2 Wiring
WN	–	Webbed Neck

PATIENT CONSENT FORM

Study Details : **A Comprehensive study on Craniovertebral Junction Anomalies**

Study Centre : **Institute of Neurology,**
Madras Medical College and
Rajiv Gandhi Government General Hospital,
Chennai - 600 003.

Patient may check (✓) these boxes:

I confirm that I have understood the purpose of procedure for the above study. I have the opportunity to ask question and all my questions and doubts have been answered to my complete satisfaction. ☐

I understand that my participation in the study is voluntary and that I am free to withdraw at any time without giving reason, without my legal rights being affected. ☐

I understand that the investigator of the clinical study, others working on his behalf, the ethical committee and the regulatory authorities will not need my permission to look at my health records, both in respect of current study and any further research that may be conducted in relation to it, even if I withdraw from the study. However, I understand that my identity will not be revealed in any information released to third parties or published, unless as required under the law. I agree not to restrict the use of any data or results that arise from this study. ☐

I agree to take part in the above study and to comply with the instructions given during the study and faithfully cooperate with the study team and to immediately inform the study staff if I suffer from any deterioration in my health or wellbeing or any unexpected or unusual symptoms. ☐

I hereby give permission to undergo complete clinical examination and diagnostic tests including hematological, biochemical, radiological, EMG, EEG, NCS, Lumbar puncture and muscle biopsy, appropriate to the clinical diagnosis. ☐

I hereby consent to participate in this study. ☐

Signature / Thumb impression: Place : Date :

Patient Name and Address:

Signature of Investigator: Place : Date
Study Investigator's Name :

சுய ஒப்புதல் படிவம்

ஆய்வு செய்யப்படும் தலைப்பு : **A Comprehensive study on Craniovertebral Junction Anomalies**

ஆராய்ச்சி நிலையம் : நரம்பியல் மருத்துவத் துறை,
இராஜீவ் காந்தி அரசு பொது மருத்துவமன
மற்றும் சென்னை மருத்துவக்கல்லூரி,
சென்னை - 600 003.

பங்கு பெறுபவரின் பெயர் : **உறவு முறை:**

பங்கு பெறுபவரின் எண் :

பங்கு பெறுபவர் இதனை (√) குறிக்கவும்

மேலே குறிப்பிட்டுள்ள மருத்துவ ஆய்வின் விவரங்கள் எனக்கு விளக்கப்பட்டது. என்னுடைய சந்தேகங்களைக் கேட்கவும், அதற்கான தகுந்த விளக்கங்களைப் பெறவும் வாய்ப்பளிக்கப்பட்டது. ☐

நான் இவ்வாய்வில் தன்னிச்சையாகத்தான் பங்கேற்கிறேன். எந்தக் காரணத்தினாலோ எந்தக் கட்டத்திலும் எந்த சட்ட சிக்கலுக்கும் உட்படாமல் நான் இவ்வாய்வில் இருந்து விலகிக் கொள்ளலாம் என்றும் அறிந்து கொண்டேன். ☐

இந்த ஆய்வு சம்மந்தமாகவும், மேலும் இது சார்ந்த ஆய்வு மேற்கொள்ளும்போதும், இந்த ஆய்வில் பங்குபெறும் மருத்துவர் என்னுடைய மருத்துவ அறிக்கைகளைப் பார்ப்பதற்கு என் அனுமதி தேவையில்லை என அறிந்துகொள்கிறேன். நான் ஆய்வில் இருந்து விலகிக் கொண்டாலும் இது பொருந்தும் என அறிகிறேன். ☐

இந்த ஆய்வின் மூலம் கிடைக்கும் தகவல்களையும், பரிசோதனை முடிவுகளையும் மற்றும் சிகிச்சை தொடர்பான தகவல்களையும் மருத்துவர் மேற்கொள்ளும் ஆய்வில் பயன்படுத்திக் கொள்ளவும், அதைப் பிரசுரிக்கவும் என் முழு மனதுடன் சம்மதிக்கிறேன். ☐

இந்த ஆய்வில் பங்கு கொள்ள ஒப்புக்கொள்கிறேன். எனக்குக் கொடுக்கப்பட்ட அறிவுரைகளின் படி நடந்துகொள்வதுடன், இந்த ஆய்வை மேற்கொள்ளும் மருத்துவ அணிக்கு உண்மையுடன் இருப்பேன் என்றும் உறுதியளிக்கிறேன். என் உடல் நலம் பாதிக்கப்பட்டாலோ அல்லது எதிர்பாராத வழக்கத்திற்கு மாறாக நோய்க்குறி தென்பட்டாலோ உடனே அதை மருத்துவ அணியிடம் தெரிவிப்பேன் என உறுதி அளிக்கிறேன். ☐

இந்த ஆய்வில் எனக்கு மருத்துவப் பரிசோதனை, இரத்தப் பரிசோதனை மற்றும் நரம்பு மின் பரி ☐

சோதனை செய்து கொள்ள நான் முழு மனதுடன் சம்மதிக்கிறேன்.

பங்கேற்பவரின் கையொப்பம் ..... இடம் ..... தேதி
கட்டைவிரல் ரேகை:

பங்கேற்பவரின் பெயர் மற்றும் விலாசம்

ஆய்வாளரின் கையொப்பம் ..... இடம் ..... தேதி

ஆய்வாளரின் பெயர் .....